2012 Hematopoietic Coding Manual
Effective with Cases Diagnosed 1/1/2012 and after

Editors: Jennifer Ruhl, RHIT, CCS, CTR, NCI SEER
Margaret (Peggy) Adamo, BS, AAS, RHIT, CTR, NCI SEER
Lois Dickie, CTR, NCI SEER

Developer /consultant: Carol Hahn Johnson, BS, CTR, Consultant

Suggested citation: Johnson CH, Adamo M, Dickie L, Percy-Laurry A, Ruhl J (eds.). 2012 Hematopoietic Manual. Version 2.2. National Cancer Institute, Bethesda, MD 20892-8316

In Appreciation

NCI SEER would like to acknowledge the dedicated work of Drs Charles Platz and Graca Dores since the inception of the Hematopoietic project. We deeply appreciate their willingness to serve as advisors for the rules within this manual. The quality of this Hematopoietic project is directly related to their commitment.

NCI SEER would also like to acknowledge the following individuals who provided substantial input on this version of the manual. Their contributions are greatly appreciated

- Cynthia Dryer (SEER Iowa State Registry)
- Tiffany Janes (SEER Seattle Regional Registry)
- Mary Mrosczyk (Maine Cancer Registry)
- Lisa Pareti (SEER Louisiana State Registry)
- Steven Peace (Florida Cancer Data System)
- Winny Roshala (SEER Greater California Registry)
- Frances Ross (SEER Kentucky Registry)
- Wendy Scharber (Registry Widgets)

Dedication

This version of the Hematopoietic Manual and the companion Hematopoietic and Lymphoid Neoplasm Database (Hematopoietic DB) are dedicated to the hard-working cancer registrars across the world that meticulously identify, abstract and code cancer data. Cancer registrars are the foundation for statewide, provincial, territorial, national, and international cancer surveillance programs which support cancer prevention and cancer control efforts worldwide.

2013 Revisions

This manual and the corresponding database are to be used for coding cases diagnosed January 1, 2012 and forward. **The changes made do not require registrars to recode old cases.**

Rule numbers are the same as those in the 2012 version. There are no new or revised rules. The revisions include

- Correction of typos (in both manual and database)
- Clarification of rules
- Examples for rules added
- Rules available in text format only (flowchart and matrix formats not available)
- A new User's Guide is available to help illustrate searching the database.

Many of the revisions were based on questions received in Ask SEER CTR.

Table of Contents

Introduction 6
Case Reportability Instructions 16
Multiple Primary Rules 18
Primary Site and Histology Coding Rules 24
Module 1: Post-Transplant Lymphoproliferative Disorder 9971/3 (PH1-PH2) 24
Module 2: Plasma Cell Neoplasms (PH3-PH7) 25
Extraosseous plasmacytoma 9734/3
Solitary plasmacytoma of bone 9731/3
Plasma cell myeloma/multiple myeloma 9732/3
Plasma cell leukemia 9732/3
Module 3: Lymphoma/Leukemia (PH8-PH10) 26
Adult T-cell leukemia/lymphoma (HTLV-1 positive) 9827/3
Adult T-cell leukemia/lymphoma 9837/3
B Lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL) 9816/3
B Lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3);E2A-PBX1 (TCF3-PBX1) 9818/3
B Lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1) 9814/3
B Lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH 9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1 9812/3
B lymphoblastic leukemia/lymphomas, NOS 9811/3, 9812/3-9818/3
Blastic natural killer leukemia/lymphoma 9727/3
Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL) 9823/3
Lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged 9813/3
Module 4: Preleukemia, Smoldering leukemia and Myelodysplastic syndrome 9989/3 (PH11) 29
Module 5: Myeloid Neoplasms and Mast Cell Neoplasms (PH12-PH15) 29
Acute myeloid leukemia, NOS 9861/3, 9811/3-9897/3
Mast Cell Leukemia 9742/3
Mast cell sarcoma 9740/3
Myeloid sarcoma 9930/3
Module 6: Coding Primary Site and Histology for Specified Lymphoma (PH16-PH24) 30
B-cell lymphoma, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma 9596/3
Diffuse large B-cell lymphoma 9680/3
Follicular lymphoma 9690/3
Follicular lymphoma, grade 1 9695/3
Follicular lymphoma, grade 2 9691/3
Follicular lymphoma, grade 3, 3A, 3B 9698/3
Lymphoplasmacytic lymphoma 9671/3
Primary cutaneous follicle center cell lymphoma 9597/3
Waldenstrom macroglobulinemia 9761/3
Module 7: Primary Site Rules for Lymphomas Only 9590/3-9729/3, 9811/3-9818/3, 9823/3, 9827/3, 9837/3 (PH25-PH37) 32

Module 8: Multiple Histologies Coded as a Single Primary: Hodgkin and non-Hodgkin Lymphoma AND Multiple non-Hodgkin Lymphomas (PH38-PH-39).. 34
Module 9: NOS and More Specific Histology: All hematopoietic and lymphoid neoplasms 9590/3-9992/3 (PH40-PH41) 35
Module 10: Coding Primary Site and Histology when Modules 1-9 are N/A: All hematopoietic and lymphoid neoplasms 9590/3-9992/3 (PH42-PH43)..... 35
Grade of Tumor Rules..... 36
Glossary 39
Appendix A History of Hematopoietic and Lymphoid Neoplasm Coding..... 51
Table A1: Histiocytic and Dendritic Cell Neoplasm Obsolete Terms..... 53
Table A2: Hodgkin Lymphoma (Hodgkin Disease) Obsolete Terms 53
Table A3: Lymphoid Neoplasm Obsolete Terms..... 54
Table A4: Myeloid Neoplasm Obsolete Terms 55
Appendix B WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues Histology Lineage 56
Table B1: Myeloproliferative Neoplasms..... 56
Table B2: Myeloid and Lymphoid Neoplasms with Eosinophilia and Abnormalities of PDGFRA, PDGFRB or FGFR1 56
Table B3: Myelodysplastic / Myeloproliferative Neoplasms 56
Table B4: Myelodysplastic Syndromes 57
Table B5: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms..... 57
Table B6: Acute Leukemia of Ambiguous Lineage 58
Table B7: Precursor Lymphoid Neoplasms..... 58
Table B8: Mature B-Cell Neoplasms..... 58
Table B9: Mature T-Cell and NK-Cell Neoplasms 59
Table B10: Hodgkin Lymphoma 60
Table B11: Histiocytic and Dendritic Cell Neoplasms..... 60
Table B12: Post-Transplant Lymphoproliferative Disorders (PTLD)..... 60
Appendix C Lymph Node/Lymph Node Chain Reference Table..... 61
Table C1: Lymph Node/Lymph Node Chain Reference Table 61
Appendix D New Histology Terms and Codes Hematopoietic and Lymphoid Neoplasms 69
Table D1a: New Histology Terms and Codes – Alphabetic List..... 69
Table D1b: New Histology Terms and Codes – Numeric List..... 70
Table D2: Histologic Terms and Codes with Changes in Case Reportability (Newly Reportable Conditions)..... 71
Appendix E: Obsolete Histologies..... 72

Introduction and Background

The Hematopoietic Working Group was led by the Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute (NCI) and included members from many professional organizations: the National Cancer Registrars Association (NCRA), the North American Association of Central Cancer Registries (NAACCR), the National Program of Cancer Registries (NPCR) of the Centers for Disease Control and Prevention (CDC), the Commission on Cancer (CoC) of the American College of Surgeons (ACoS), and the Canadian Cancer Registries (CCR). The Working Group also included cancer registrars who work independently (contractors), hospital registrars, central cancer registry registrars, and clinical and research physicians who are experts in the hematopoietic and lymphoid neoplasm fields.

This working group has developed rules, guidelines and an interactive desktop Hematopoietic and Lymphoid Database (Hematopoietic DB) reference to assist registrars in determining case reportability, the number of primaries, as well as instructions for coding primary site, histology, and grade for a hematopoietic and/or lymphoid neoplasm. The rules, guidelines, and the Hematopoietic DB follow the *World Health Organization (WHO) Classification of Tumours of the Haematopoietic and Lymphoid Tissues*, 4th Edition, 2008, also called the "WHO Blue Book." Both the *International Classification of Diseases for Oncology (ICD-O)* and the series of Blue Books are produced by the World Health Organization (WHO), but the content of the books are very different. Each has a prominent place in the registry world.

The original ICD-O, the ICD-O-2, and the ICD-O-3 provide standard primary site and histology codes for specific benign, borderline, and malignant conditions. The ICD-O series also provides generic "not otherwise specified" or "NOS" codes for some conditions so registrars are able to code cases that have limited information, such as death-certificate-only cases and historic cases. When ICD-O attributes a code to a specific histology, the original code is rarely changed. The intent is that the code should never change; for example, code 8140/3 for adenocarcinoma, NOS has remained unchanged since the first edition of ICD-O. The ICD-O manuals are the standard for coding neoplasms throughout the world. To preserve the integrity of historical data, and to allow for comparison of data over time, it is imperative that standard codes remain unchanged. Although the stability of these codes is necessary to interpret data over time, that process has some less-than-desirable results. When the clinical world reclassifies diseases to reflect the current state of science and knowledge about a particular disease or condition, that disease will remain in the same numeric position in ICD-O. When the ICD-O editors assign new codes for a neoplasm, the new code may not be placed in the desired category because there may not be room within that category to add a new code. An example of this problem is the placement of the non-Hodgkin lymphomas that were first added in ICD-O-3.

The WHO Blue Books, by contrast, are histo-pathology reference books used by pathologists and oncologists throughout the world. The Blue Books are revised and published when new information is available. The *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition, a collaborative project of the Society for Hematopathology/European Association for Hematopathology was published in 2008. The reference includes new disease classifications, changes to existing classifications and cell lineages, and new conditions that reflect the state-of-the-science for these neoplasms. This reference was the primary source of information used to develop the *2010 Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual*, the *2012 Hematopoietic Coding Manual*, and the accompanying Hematopoietic DB because the WHO Blue Book is consistently updated with the current classification by cell lines or lineages and classification groupings. Using the WHO classifications gives the registrar reference material that is clinically relevant and compatible with current pathology reports and medical records. When the clinical field finds specific tumor markers, immunohistochemical testing, genetic testing, or other characteristics that define or refine a diagnosis or a particular histology, the WHO Blue Books introduce proposed new codes for new or more specific histologies, and these new histologies may be grouped or classified in categories based on information about the phenotype or behavior of the neoplasm.

Note: The WHO is the directing and coordinating authority for health within the United Nations system. It is responsible for providing leadership on global health matters, shaping the health research agenda, setting norms and standards, articulating evidence-based policy options, providing technical support to countries and monitoring and assessing health trends. (From http://www.who.int/about/en/)

The 2012 *Hematopoietic Coding Manual* and the Hematopoietic DB are designed to help the registrar understand and interpret the information written by pathologists and clinicians. The Hematopoietic DB will be updated as needed to ensure that the registrar has the most current information available to interpret and code a hematopoietic or lymphoid neoplasm. **Please see http://seer.cancer.gov/tools/heme/revisions.html for the revision history.**

Leukemias and Lymphomas

Leukemia vs. Lymphoma

One of the differences between leukemia and lymphoma is that leukemia most commonly presents in the bone marrow and/or blood while lymphoma most commonly manifests in lymph nodes, lymphoid tissue, or lymphoid organs. When only the bone marrow is involved, the histology is usually leukemia. Although rare, a lymphoma may present only in the bone marrow (See Module 7 for instructions on coding primary site for lymphomas).

Both leukemia and lymphoma patients may have splenomegaly (enlargement of the spleen). Patients with leukemia may have leukemic infiltrate of the spleen. Splenomegaly does not mean that the leukemia originated in the spleen or that this neoplasm is lymphoma. The spleen filters and stores blood cells. The spleen involvement is usually secondary, much like metastases in solid tumors. The rare histologies that are primary in the spleen are identified in the Hematopoietic DB. The Primary Site will be listed as C422.

Diagnostic Process for Leukemia

For most patients, the first suspicion or presentation of a hematopoietic neoplasm will be symptoms such as unexplained weight loss, weakness, chronic fatigue, easy bruising, etc. When the physician suspects leukemia, he/she usually orders a complete blood count (CBC) and/or a peripheral blood smear. The CBC and/or peripheral smear will identify abnormalities of the platelets, hemoglobin, white blood cells or the red blood cells. When an abnormality is identified in the blood cell analysis, a bone marrow (BM) biopsy is usually the next procedure. The CBC and bone marrow seldom provide a definitive diagnosis; however, the results usually provide provisional diagnoses such as: myeloproliferative neoplasms, myeloid neoplasms, myelodysplastic/myeloproliferative neoplasms, myelodysplastic syndromes, or leukemia. These diagnoses are differential or provisional. More testing is needed to identify the specific hematopoietic or lymphoid neoplasm and subsequent treatment. Many of the neoplasms in the 2008 WHO classification require immunophenotyping or genetic information to identify the specific histology. The Hematopoietic DB contains information on the types of diagnostic tests that are used to identify the specific histology for the hematopoietic or lymphoid neoplasm being abstracted. See the "Definitive Diagnostic Method" section in the Hematopoietic DB.

Information for Lymphoma Only

Biopsies

The most accessible involved lymph node or site is usually biopsied when lymphoma is suspected. For example, if a CT or PET scan identified enlarged cervical and mediastinal lymph nodes, the physician would biopsy the cervical lymph nodes because that would be the least invasive procedure; i.e. the cervical nodes are more accessible than the mediastinal nodes. Do **not** assume that the more accessible site is the primary site. Follow the primary site rules and instructions.

Hodgkin Lymphoma

Hodgkin lymphoma (HL) is a type of lymphoma originating in lymphocytes (a type of white blood cell). HL is characterized by the presence of Reed-Sternberg cells (RS cells) on microscopic examination. HL usually originates in the lymph nodes and is characterized by the orderly spread of neoplasm from one lymph node chain to another. The neoplasm may progress to involve the spleen, liver, and/or bone marrow.

Non-Hodgkin Lymphoma

Non-Hodgkin lymphoma (NHL) comprises a diverse group of malignant neoplasms which include all lymphomas other than Hodgkin. NHL arises in lymphocytes (a type of white blood cell). Lymphocytes are present in lymph nodes and throughout the body. NHL occurs in extranodal sites including: tonsils, spleen, ileum, stomach, Waldeyer ring, bone marrow, skin, bone, central nervous system, lung, gonads, conjunctiva, ocular adnexa, liver, kidneys, and uterus.

The Hematopoietic Database

The Hematopoietic DB (Heme DB) is available online through the SEER website. Access the database at http://seer.cancer.gov/seertools/hemelymph/. Please note that the online version cannot be downloaded onto a PC or laptop. An internet connection is required and you must use the Heme DB online.

There is also a stand-alone version of the DB that can be downloaded onto PCs and laptops. The two versions of the Hematopoietic Database are provided currently, but future plans are to provide only the online version.

The Heme DB enables registrars to identify and understand hematopoietic and lymphoid neoplasms as well as to correctly and consistently abstract and code cases. Users are able to query any final, differential, or provisional diagnosis in the Heme DB. The diagnostic or confirmatory tests are listed under "Definitive Diagnostic Methods" for each neoplasm. The information needed to search the medical record for specific diagnostic test results is provided. Some healthcare institutions may "file" confirmatory test results, such as immunophenotyping or genetic testing, in a location other than the location used for standard laboratory tests in the medical record. We recommend that the registrar ask the laboratory for examples of test results, such as immunophenotyping or genetic testing, to become familiar with the format of the test results as well as other information that may be included with test results. We also recommend that the registrar ask the Health Information Management or Medical Records Department where these tests are "filed" within the chart (paper or electronic).

Coding the Data Item Diagnostic Confirmation

Note: Codes 1-4 are microscopically confirmed. Codes 5-8 are **not** microscopically confirmed

Code 1: Positive histology

1. For **all hematopoietic** neoplasms, positive histology includes: Biopsy of bone marrow, lymph node(s), organ(s), or any other tissue(s).
   Note: Specimens may be from a biopsy, surgery, or autopsy
2. For **leukemia only**: In addition to the procedures listed in #1, positive histology also includes CBC and peripheral blood smear.
   - Use Code 1 when the neoplasm is microscopically confirmed (definitions 1 or 2 above) **AND**
     - **No** immunophenotyping, genetic testing or JAK2 are done

     OR
     - Immunophenotyping, genetic testing or JAK2 are done but are **negative** for the **disease being abstracted**

     Example: Bone marrow is suspicious for myeloproliferative neoplasm, probably essential thrombocythemia. JAK2 was negative. Use code 1. The JAK2 did not confirm the disease.

     OR
     - Immunophenotyping, genetic testing or JAK 2 are done but are **not listed** in the Definitive Diagnostic Methods in the Hematopoietic DB

     Note: The immunophenotyping, genetic testing or JAK2 may have been done to **rule out** other neoplasms that are clonally similar to the disease being abstracted. Usually the provisional diagnosis will be two or more diseases.

Code 2: Positive cytology

- Rarely used for hematopoietic neoplasms. May be used when the diagnosis is based on

i. Examination of fluid such as spinal fluid, peritoneal fluid, or pleural fluid
ii. Paraffin block specimens from concentrated spinal fluid, peritoneal fluid, or pleural fluid
iii. When a small-gauge needle is used to obtain a specimen and there is not enough tissue to do a histologic examination. The report will be a cytology report rather than a pathology report.

Code 3: Positive histology PLUS positive immunophenotyping or genetic testing

- Use Code 3 when there is histologic confirmation (i. below) **and** the testing is listed in the Definitive Diagnostic Methods in the Hematopoietic DB (ii.) **plus either** iii. or iv. below

i. A bone marrow or tissue biopsy (or CBC or peripheral smear for leukemias only) is
   - **Positive** for the neoplasm being abstracted **OR**
   - **Suspicious (or any term on the ambiguous list)** for the neoplasm being abstracted **OR**
   - Provided a **provisional diagnosis** of the disease being abstracted **AND**

ii. Immunophenotyping, genetic testing or JAK2 are listed in the Definitive Diagnostic Methods in the Hematopoietic DB **AND**
iii. The immunophenotyping, genetic testing or JAK2 is **positive** for the disease being abstracted (confirmed the disease) **OR**
iv. The immunophenotyping, genetic testing, or JAK2 identified a **more specific histology**

Example 1 (Identifying a more specific histology): Bone marrow biopsy is positive for acute myeloid leukemia 9861/3. Genetic testing shows AML with inv(16)(p13.1q22) 9871/3. Code the Diagnostic Confirmation 3, positive histology and positive genetic testing.

Example 2 (Identifying a more specific histology): Skin biopsy is positive for cutaneous T-cell lymphoma, NOS 9709/3. Immunophenotyping shows CD8 positive. Diagnosis is primary cutaneous CD8 positive aggressive epidermitropic T-cell lymphoma 9709/3. Code the Diagnostic Confirmation 3, positive histology and positive genetic testing.

Example 3 (Confirming the histologic diagnosis): The diagnosis from the bone marrow biopsy is plasma cell dyscrasia. Peripheral blood smear is compatible with plasma cell leukemia. FISH and chromosome analysis revealed plasma cell myeloma. Both plasma cell leukemia and plasma cell myeloma are coded to the

same ICD-O-3 code, so there is only one disease process. The peripheral blood smear is histologic diagnosis for the plasma cell leukemia and FISH confirmed the diagnosis of multiple myeloma/plasma cell myeloma. Code the Diagnostic Confirmation 3, positive histology and positive genetic testing.

Example 4 (Histologic confirmation plus genetic and immunophenotyping confirmation): Patient diagnosed with CLL by CBC and flow cytometry that was positive for both the genetic and CD antigens (immunophenotyping) for CLL. A bone marrow biopsy not performed. Since this is leukemia, the CBC is histologic confirmation, so this patient had histologic confirmation, genetic, and immunophenotyping positive for CLL. Code the Diagnostic Confirmation 3, positive histology and positive genetic testing.

Code 4: Positive microscopic confirmation, method not specified

- Rarely used for hematopoietic neoplasms

Code 5: Positive laboratory test/marker study

- This code would be rarely, if ever, used for hematopoietic neoplasms. If there was no provisional diagnosis or clinical suspicion of cancer, immunophenotyping or genetic testing would not be done.

Code 6: Direct visualization without microscopic confirmation

- This code would rarely be used for hematopoietic neoplasms. The code would be used when
  - The operative report states the patient had lymphoma, but no biopsy or cytology was done
  - Gross autopsy finding of hematopoietic disease, but no tissue or fluid were examined (histology or cytology)

Code 7: Radiology and other imaging techniques without microscopic confirmation

- This code would rarely be used for hematopoietic neoplasms
- Example of a valid code 7 would be a terminally ill patient who has a CT scan with the impression: Suspicious for lymphoma. The patient refused further workup.

Code 8: Clinical diagnosis only (other than 5, 6, or 7)

Note 1: While clinical diagnosis is seldom used for solid tumors, it is a valid diagnostic method for certain hematopoietic neoplasms. The Hematopoietic DB will list Clinical Diagnosis as the definitive diagnostic method for these neoplasms.

Note 2: For these neoplasms, the biopsy, immunophenotyping, and genetic testing do not confirm the neoplasm. The diagnosis is determined based on the physician's clinical expertise, combined with the information from the biopsy, equivocal or negative tests, and the clinical symptoms. This is called a "diagnosis of exclusion" because the physician's judgment and the work-up literally exclude all other possible diagnoses, leaving one diagnosis.

Code 9: Unknown whether or not microscopically confirmed; death certificate only

- Use when
  - There is minimal information on the case; diagnostic confirmation method is not known
  - Death certificate only cases

Transformation: Chronic Neoplasm to Acute Neoplasm (Rules M10-M15)

A chronic neoplasm is a neoplasm that can transform to an acute/more severe neoplasm; the Hematopoietic DB will show the acute neoplasm in the "Transformations" section. For example, if you search the Hematopoietic DB for chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL), the "Transformations" section shows that CLL/SLL transforms to diffuse large B-cell lymphoma and Hodgkin lymphoma. That means that CLL/SLL is a chronic neoplasm and both diffuse large B-cell lymphoma and Hodgkin lymphoma are acute neoplasms.

The most common transformation is when a neoplasm progresses from chronic to acute. However, neoplasms may be diagnosed in an acute phase and transform to the less aggressive chronic phase after treatment. Rules M14 and M15 cover this type of situation.

Rule M15 says if the acute neoplasm is diagnosed first and the chronic is diagnosed more than 21 days later, it is important to determine if the patient received treatment for the acute neoplasm. If the patient was treated, abstract the chronic neoplasm as a second primary.

If the patient was **not** treated for the acute neoplasm, use Rule M14; code a single primary, the acute neoplasm.
Note: Follow back to determine whether there was any further diagnostic workup that proved the acute diagnosis was incorrect or documentation that the acute diagnosis was provisional.

Examples of acute and chronic diseases (Rules M10-M15):

1. Rule M10: Pathology states 20% myeloblasts consistent with high grade myelodysplastic syndrome evolving into acute leukemia. Rule M10 says this is a single primary since both the chronic and the acute neoplasm are diagnosed **at the same time** with only **one positive biopsy**.
2. Rule M12: Consult from outside pathology date July 26, 2013 states AML arising from known MDS. Previous MDS diagnosed August 23, 2010. Abstract as two primaries. When determining if you have one or two primaries, you go by the date of diagnosis of the second diagnosis. There is a chronic neoplasm (the MDS) and an acute neoplasm (the AML) diagnosed more than 21 days apart.
3. Rule M13: Two pathology reports. Bone marrow on 6/23/2013 revealed lymphoplasmacytic lymphoma. On 6/24/2013 right cervical lymph node biopsy shows diffuse large b-cell lymphoma (DLBCL). Code as two primaries, lymphoplasmacytic lymphoma and DLBCL. The Transformations section in the Hematopoietic DB says that lymphoplasmacytic leukemia (chronic) transforms to DLBCL (acute). Since you have two biopsies, a bone marrow biopsy confirming the lymphoplasmacytic lymphoma (chronic disease) and a lymph node biopsy confirming the DLBCL (acute disease), Rule M13 applies and you code this as two primaries.

Obsolete Hematopoietic Histology Codes

Defining obsolete codes in the hematopoietic database and manual

As new classifications for hematopoietic neoplasms are developed, certain codes may be determined to be "obsolete" or no longer used. The Hematopoietic DB is based on the 2008 "*WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues"(2008 WHO)*. The 2008 WHO reclassified some terms based on the most recent scientific understanding of these neoplasms. The 2008 WHO reclassification rendered some codes obsolete and introduced new codes. Terms associated with obsolete codes have been reclassified in the 2008 WHO by moving them to another existing code (ICD-O-3) or to one of the new codes.

Most of the codes that became obsolete based on the 2008 WHO became obsolete for cancer registry data collection as of 1/1/2010 with the implementation of the 2010 Hematopoietic database and manual. Additional codes became obsolete for data collection as of 1/1/2012. The codes that became obsolete based on the 2008 WHO are identified by "[OBS]" in the Hematopoietic DB. The current code to be used in place of the obsolete code is shown in the database. These codes will continue to appear in ICD-O-3, so it is important for registrars to use the Hematopoietic Database and Manual to determine the correct histology code. The codes will also continue to appear in cancer reporting software, legacy data grouping and data reports.

Note: These instructions for coding [OBS] codes are for hematopoietic neoplasms (9590-9992) only. For all other histologies, refer to ICD-O-3 for coding instructions.

When to use an obsolete code

Obsolete [OBS] codes in the database will have the following instruction: "This code can be used for cases diagnosed prior to 1/1/2010 (or 1/1/2012), DCO cases, and cases with minimal information." The use of these codes is dependent on the **date of diagnosis**.

Cases diagnosed prior to 2010 (or 2012)

For cases that are diagnosed prior to the effective obsolete date, use the OBSOLETE code.

Example 1: Pathology report dated 7/12/2009 states therapy related myelodysplastic syndrome. Code histology to 9987/3 since the date of diagnosis is prior to 1/1/2010. (Histology code 9987/3 became obsolete 1/1/2010).

Example 2: Pathology report dated 12/15/2011 states malignant lymphoma, large B-cell, diffuse immunoblastic, NOS. Code histology to 9684/3 since the date of diagnosis is prior to 1/1/2012. (Histology code 9684/3 became obsolete as of 1/1/2012)

Death Certificate Only (DCO) cases

For death certificate cases where a date of diagnosis can be determined through follow back, use that date. If there is a diagnosis that matches one of the obsolete codes, determine when the code became obsolete and assign the appropriate histology code.

Example 1*:* Death certificate states plasma cell leukemia (9733/3). Date of death is 10/6/2012. Through follow back, patient found to have been diagnosed in 2011. Code the histology to Multiple Myeloma (9732/3). Plasma cell leukemia (9733/3) became obsolete as of 1/1/2010 and the heme db lists 9732 as the current code.

Example 2*:* Death certificate states Hodgkin lymphoma, nodular sclerosis, grade 1 (9665/3). Date of death is 7/18/2012. Through follow back, date of diagnosis is determined to be "3-4 years ago." Since this case was diagnosed prior to 2010, assign histology code 9665/3.

Example 3*:* Death certificate states small lymphocytic lymphoma (9670/3). Date of death is 12/8/2012. Through follow back, date of diagnosis is determined to be "a few months ago." Since this case was diagnosed after 1/1/2012, assign histology code 9823/3. 9670/3 became obsolete as of 1/1/2012.

Example 4*:* Death certificate states malignant histiocytosis (9750/3). Date of death is 3/15/2012. Per the Hematopoietic database, code 9750/3 became obsolete as of 1/1/2010. Additional information on date of diagnosis cannot be determined. Code the histology to 9750/3 even though the date of diagnosis (date of death) is after 1/1/2010 because this is a DCO case and the date of diagnosis is unknown.

Cases with minimal information

In situations where limited information is available, i.e., path only case, with no date of original diagnosis, use the following instructions.

- If no date of diagnosis can be determined, use the date on the pathology or cytology report and the current code.

  Example: Path report from liver biopsy on 6/23/2012 shows malignant lymphoma, large B-cell, diffuse, immunoblastic (9684/3). Reason for biopsy states "h/o lymphoma." No other information regarding diagnosis. Code the histology to 9684/3.

- If an original date of diagnosis can be determined, then use the instructions for that date.

  Example*:* Path report from liver biopsy on 6/23/2012 shows malignant lymphoma, large B-cell diffuse, immunoblastic (9684/3). Further review of patient's records shows that patient was originally diagnosed in 2006 with large B-cell diffuse, immunoblastic (9684/3) in the stomach. Code the histology to 9684/3.

See Appendix E for a listing of the obsolete codes.

Steps in Priority Order for Using the Heme DB and Hematopoietic Coding Manual

1. Search the Heme DB using a unique word in the diagnosis.
    A. You can search on the complete name (diagnosis). For example, if the diagnosis is acute myelomonocytic leukemia, search on the complete term, acute myelomonocytic leukemia. The number of matched terms that are displayed will be much smaller.
        I. The search engine will display every entry with **all** of the words "acute, myelomonocytic, and leukemia." The results displayed will have all three words in the histology name.
    B. You can also search on abbreviations such as AMML for acute myelomonocytic leukemia, DLBCL for diffuse large B-cell lymphoma, or AML for acute myeloid leukemia.
2. When multiple results are displayed, click on the selected term (acute myelomonocytic leukemia) to display the record.

3. Use the displayed record to
    A. Determine the correct **histology code.**
    B. Determine **primary site.**
        I. The primary site code displayed under **Primary Site(s)** is the **only** site code to be used for that histology
            a. For certain primaries, only one primary site code is displayed. There are no exceptions for leukemia, MDS, or MPD.
            Note 1: All leukemias are assigned primary site bone marrow C421. There are no exceptions. This rule was implemented in ICD-O-2 in 1992.
            Note 2: For lymphoma only, the following are examples of exceptions
                i. When lymphoma is present only in the bone marrow, code the primary site to bone marrow
                ii. When lymphoma is present only in a site not listed under **Primary Site(s)**, code to the site of origin.
                iii. When the physician states that the lymphoma originated in a site other than the site listed under **Primary Site(s)**, code to the site of origin using the physician's statement
            b. When there is no primary site code listed under **Primary Site(s)**, read the **Abstractor Notes**. The Abstractor Notes will contain information on the most common primary sites, less common primary sites, and other sites of involvement for stages II, III, and IV lymphomas. Use the Abstractor Notes to confirm that the **site/histology combination is probable.**
            c. Search the Hematopoietic Manual to find applicable Multiple Primary (M), Primary Site and Histology (PH), and Grade (G) rules.
            Note: Search efficiently by entering the histology code obtained from the Heme DB into the Adobe search function to quickly move through the rules.
            d. When there is no primary site code listed under **Primary Site(s)**, read the abstractor notes to find the common and less common sites for that particular neoplasm.
4. Hematopoietic Manual Special Modules; When and how to use:
    A. **Module 7**: Coding Primary Site for Lymphomas Only; Use when:
        I. Assigning a primary site is difficult
        II. Multiple lymph node chains are involved
        III. Organ(s) and lymph nodes are involved
        IV. Lymphoma presents as a mass in mediastinum, etc.
        V. Unknown primary site
    B. **Module 8**: Multiple Histologies Coded as a Single Primary: Hodgkin and non-Hodgkin Lymphoma AND Multiple non-Hodgkin Lymphomas
    C. **Module 9**: NOS and More Specific Histology: All Hematopoietic and Lymphoid Neoplasms
    D. **Module 10**: Coding Primary Site and Histology. Use Only When Modules 1-9 are Not Applicable

5. Use the Hematopoietic Multiple Primaries Calculator only when instructed by the rules in the Hematopoietic Manual.

Reporting Phlebotomy, Blood-Thinners/Anti-Clotting Medications, and Transfusions as Treatment

- Do **not** collect **blood transfusions** (whole blood, platelets, stem cell transplants etc.) as treatment. Blood transfusions are used widely to treat anemia and it is not possible to collect this procedure in a meaningful way.
  Note: This is a new instruction for cases diagnosed 1/1/2012 and later

- Collect **phlebotomy** for polycythemia vera ONLY.
  Note: This is an addition to the 2010 instructions.

- Collect **blood-thinners** and/or **anti-clotting agents** for
  9740/3 Mast cell sarcoma
  9741/3 Systemic mastocytosis
  9742/3 Mast cell leukemia
  9875/3 Chronic myelogenous leukemia BCR/ABL1 positive
  9950/3 Polycythemia vera
  9961/3 Primary myelofibrosis
  9962/3 Essential thrombocythemia
  9963/3 Chronic neutrophilic leukemia
  9975/3 Myelodysplastic/myeloproliferative neoplasm, unclassifiable

Note: This information was added in the 2012 Hematopoietic manual. It is not listed in the 2010 Hematopoietic manual but can be applied to cases diagnosed 2010+.

Case Reportability Instructions

Note: In many cases, the registrar will need to make inquiries to the physician's office to confirm the diagnosis. Unless that type of follow-back is done, hematopoietic cases will be under-reported.

1. Query the **Heme DB** to determine case reportability.

2. Report all cases with morphology codes **9590-9992** with a **/3** behavior.
   Note: In ICD-O-3 preleukemia is listed as 9989/3 in the numeric list and 9989/1 in the alphabetic list. Change the 9989/1 in the alphabetic list to a 9989/3 in ICD-O-3.

3. Report hematopoietic and lymphoid neoplasms with ICD-O-3 morphology codes **9590-9992** that are listed as **/1** and **described as malignant** by a physician.
   Note: Do **not** report in situ (/2) lymphomas

4. Report the case when the diagnosis of a hematopoietic neoplasm is preceded by one of the following **ambiguous terms:**
   - Appears
   - Comparable with
   - Compatible with
   - Consistent with
   - Favor(s)
   - Malignant appearing
   - Most likely
   - Presumed
   - Probable
   - Suspect(ed)
   - Suspicious (for)
   - Typical (of)

   Note 1: Use these terms when screening all diagnoses other than cytology and tumor markers.
   Note2: Report cases that use only the words on the list or an equivalent word such as "favored" rather than "favor(s) or "likely" rather than "most likely"". Do not substitute synonyms such as "supposed" for "presumed" or "equal" for "comparable."
   Note 3: Accept the reportable term and report the case when one part of the medical record uses a reportable ambiguous term such as "apparently" and another section of the medical record(s) uses a term that is not on the reportable list.
   Note 4: Diagnoses based on ambiguous terminology require follow-back to see if the diagnosis has been confirmed or proven to be incorrect (see note 5).
   Note 5: Do **not** report the case when biopsy or physician's statement proves the ambiguous diagnosis is **wrong** (for example, pathology diagnosis is benign or borderline)
   Example: CT scan shows enlarged lymph nodes suspicious for lymphoma. Subsequent biopsies of the lymph nodes thought to be involved with a neoplasm are negative for malignancy. The pathology is more reliable than the scan; the negative biopsy proves that the ambiguous diagnosis was wrong. Do **not** report the case.
   Note 6: Do **not** report cases diagnosed only by ambiguous **cytology** (cytology diagnosis preceded by ambiguous term)
   Example: Parotid ultrasound guided FNA: consistent with Non-Hodgkin's lymphoma. This case was diagnosed based on cytology/fine needle aspiration (FNA) preceded by ambiguous terminology (**consistent with**). Do not report this case based on ambiguous cytology.

5. Report the case when the patient received all or part of their first course **treatment** at the reporting facility.
   Note 1: Report the case even if the diagnostic tests are inconclusive, equivocal, or negative
   Note 2: For treatment information see the National Cancer Institute's Physicians' Data Query (PDQ) website at http://www.cancer.gov/cancertopics/pdq/cancerdatabase or the SEER*Rx Database

6. Report the case when there is a **clinical diagnosis** (physician's statement) of reportable hematopoietic or lymphoid neoplasm.
   Note 1: The clinical diagnosis may be a final diagnosis found within the medical record or recorded on a scan (CT, MRI for example)
   Note 2: Report the case even if the diagnostic tests are equivocal. A number of hematopoietic neoplasms are "diagnoses of exclusion" in which the diagnostic tests are equivocal and the physician makes the clinical diagnosis based on the equivocal tests and the clinical picture. See the Heme DB for definitive diagnostic methods for the specific neoplasm being abstracted.

7. Report the case when a reportable diagnosis appears in any text or report described as a **definitive diagnostic method** in the Heme DB.
   Note 1: Definitive diagnostic methods differ depending upon the histology. See the Heme DB for details.
   Note 2: The instructions for reporting preleukemia, early multiple myeloma and smoldering multiple myeloma were removed from the reportability list. Those terms **are** in the Heme DB.

Multiple Primary Rules

Note 1: Within these rules, the term "chronic neoplasm" means a neoplasm that transforms to another, more acute neoplasm. When you display the chronic neoplasm in the Heme DB, the Transformations section will show the "acute neoplasm." For example, if you search the Heme DB for essential thrombocythemia, acute myeloid leukemia will be listed under Transformations. Essential thrombocythemia is the chronic neoplasm and acute myeloid leukemia is the acute neoplasm.

Note 2: The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology may be identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up.

Rule M1 Abstract a single primary* when **minimal information** is available (such as a death certificate only (DCO) case or a pathology-report-only case).

Rule M2 Abstract a single primary* when there is a **single histology.**

Note 1: Bilateral involvement of lymph nodes and/or organs is still a single primary.

Note 2: Recurrence of the same histology is always the same primary (timing is not relevant).

Note 3: A single histology is the histology diagnosed by the definitive diagnostic method as defined in the Heme DB. For example, the patient had several provisional diagnoses but the definitive diagnostic method identified a single histology. Abstract as a single primary.

Example 1: The diagnosis is multiple myeloma 9732/3. Abstract as a single primary.

Example 2: Multiple extraosseous plasmacytomas 9734/3 are present in the oropharynx. Abstract as a single primary.

Example 3: A single histology diagnosed by the definitive diagnostic method as defined in the Heme DB. For example, the patient had several provisional diagnoses but the definitive diagnostic method identifies a single histology. Abstract as a single primary.

Rule M3 For the following neoplasms, abstract a single primary* when a sarcoma is diagnosed either **simultaneously** or **after** a leukemia of the same lineage.

- Mast cell sarcoma diagnosed simultaneously with mast cell leukemia or after mast cell leukemia
- Myeloid sarcoma diagnosed simultaneously with acute myeloid leukemia/myelomonocytic leukemia or diagnosed after acute myeloid leukemia/myelomonocytic leukemia

Note: The sarcoma is a solid manifestation of the leukemia. For example, when a patient has acute myeloid leukemia, the myeloid sarcoma is the result of myeloid cells migrating from the bone marrow or blood into tissue. It is part of the disease process for the acute leukemia.

Example: Acute myeloid leukemia (AML) diagnosed in 2012. In 2013, a soft tissue mass was biopsied and the pathology report final diagnosis was myeloid sarcoma. The myeloid sarcoma is a manifestation of the AML. The malignant myeloid cells are present in the blood. One of the malignant myeloid cells lodged in a capillary and grew in the tissue forming a myeloid cell soft tissue mass (referred to as myeloid sarcoma). This is not a second primary, it is a direct result of the myeloid cells circulating in the blood. It is not unlike a solid tumor in the colon metastasizing to the liver.

Rule M4 Abstract a single primary* when a **specific MDS subtype** is followed by a **different specific MDS subtype.**

Note 1: MDS is a group name for a number of specific diseases. As the MDS progresses, it may manifest as several subtypes. This is a part of the disease process. Abstracting each of the subtypes would result in over-counting this particular disease.

Note 2: Code the first subtype that is diagnosed. Do not change the histology code or create a new abstract for any subsequent specific MDS subtypes.

Note 3: Find the MDS subtypes in the Heme DB Abstractor Notes for MDS, unclassified (see code 9989/3)

* Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

Rule M5 Abstract a single primary* when both **follicular lymphoma (FL) and diffuse large B-cell lymphoma (DLBCL)** are simultaneously present in the same anatomic location(s). FL and DLBCL may be present in:

- Same lymph node(s)
- Same lymph node region(s)
- Same organ(s)
- Same tissue(s)

Note 1: Do **NOT** query the Heme DB Multiple Primaries Calculator when FL and DLBCL are present in the same site(s).

Note 2: This rule should be applied for all follicular lymphomas: FL NOS 9690/3, FL grade 1 9695/3, FL grade 2 9691/3, and FL grade 3 9698/3.

Rule M6 Abstract a single primary* when **two or more types of non-Hodgkin lymphoma** are **simultaneously** present in the **same anatomic location(s)**, such as the:

- Same lymph node(s)
- Same lymph node region(s)
- Same organ(s)
- Same tissue(s)

Note 1: Do **NOT** use this rule for cutaneous lymphomas. Simultaneous occurrences of two or more cutaneous lymphomas, other than an NOS and more specific, are extremely rare. If there are simultaneous cutaneous lymphomas, **DO NOT** use this rule and proceed to rule M16. Use the Multiple Primaries Calculator to determine whether the lymphomas are the same or different primaries.

Note 2: When the neoplasm is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for help identifying lymph node names, chains, regions and codes.

Note 3: When the neoplasm is in a more advanced stage, both non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3, or in an organ and that organ's regional lymph nodes, or in multiple organs.

- Although the combination of two or more types of non-Hodgkin lymphoma must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination of two or more types of non-Hodgkin lymphoma, assume that all of the nodes, tissue, and/or organs are involved with the same combination of non-Hodgkin lymphomas.

Note 4: Do **not** query the Heme DB Multiple Primaries Calculator in this situation.

Example: Biopsy of cervical lymph node shows **two** different non-Hodgkin lymphomas. Abstract as a single primary.

Rule M7 Abstract a single primary* when both **Hodgkin and non-Hodgkin lymphoma** are simultaneously present in the **same anatomic location(s)**. Hodgkin and non-Hodgkin may be present in the same lymph node or same lymph node region(s), the same organ(s), and/or the same tissue(s).

Note 1: When the neoplasm is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for help identifying lymph node names, chains, regions and codes.

Note 2: When the neoplasm is in a more advanced stage, both Hodgkin and non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3, or in an organ and that organ's regional lymph nodes, or in multiple organs.

- Although both Hodgkin and non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination Hodgkin and non-Hodgkin lymphomas, assume that all of the nodes, tissue, and/or organs are involved with the combination of Hodgkin and non-Hodgkin lymphomas.

Note 3: Do **not** query the Heme DB Multiple Primaries Calculator in this situation.

Example: Biopsy of cervical lymph node shows Hodgkin and non-Hodgkin lymphomas. Abstract as a single primary.

* Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

Rule M8 Abstract as multiple primaries** when **Hodgkin lymphoma** is diagnosed in one site and **non-Hodgkin lymphoma** is diagnosed in another site(s).

Example 1: Patient is diagnosed with Hodgkin lymphoma in the cervical lymph nodes and also with non-Hodgkin lymphoma in the GI tract. Abstract as multiple primaries.

Example 2: Hodgkin lymphoma in a mediastinal mass and non-Hodgkin lymphoma in the tonsil. Abstract as multiple primaries.

Rule M9 Abstract a single primary* when a **more specific** histology is diagnosed after an **NOS** ONLY when the Heme DB Multiple Primaries Calculator confirms that the NOS and the more specific histology are the same primary.

Note 1: There are no time restrictions on these diagnoses; the interval between the NOS and the more specific histology does not affect this rule stating that the **two** neoplasms are a single primary

Note 2: The Heme DB Multiple Primaries Calculator will identify these histologies as a single primary

Note 3: Change the histology code on the original abstract to the more specific histology. Check previous editions of ICD-O (i.e. ICD-O-1, ICD-O-2) to determine if the histology code was defined.

Example 1: Patient diagnosed with non Hodgkin lymphoma 9591/3 in 1999. Patient returns in 2013 with a diagnosis of CD30 positive lymphoproliferative disorder 9718/3. Histology 9591 is an NOS category and 9718 is more specific. Per the multiple primaries calculator, 9591 and 9718 are the same primary. 9718/3 was a valid code in 1999, change the histology to 9718/3 for the 1999 diagnosis.

Example 2: CT guided core biopsy pelvic mass positive for lymphoma (9591). Mediastinoscopy with biopsy shows follicular lymphoma, grade 1(9695). Lymphoma is an NOS term and follicular lymphoma is a more specific term, and they are a single primary according to the multiple primaries calculator. Abstract one primary, code the histology follicular lymphoma, grade 1 9695.

Rule M10 Abstract the **acute neoplasm** as a single primary* when both a **chronic** and an **acute** neoplasm are diagnosed **simultaneously AND** there is documentation of **only one** positive bone marrow biopsy, lymph node biopsy, or tissue biopsy.

Note: When these diagnoses happen **within 21 days**, it is most likely that one diagnosis was provisional and the bone marrow identified the correct diagnosis. Abstract the acute neoplasm.

Example: Bone marrow biopsy states MDS evolving into acute myeloid leukemia. The diagnosis of AML may be made when there is an increase in blasts but the number of blasts is less than 20%. Code to the MDS, 9989/3.

Rule M11 Abstract a single primary* and code the later histology when both a **chronic** and an **acute** neoplasm are diagnosed **simultaneously** or **less than or equal to 21 days apart AND** there is **no available documentation** on bone marrow biopsy, lymph node biopsy, or tissue biopsy.

Note 1: The two diagnoses are likely the result of an ongoing diagnostic work-up. The later diagnosis is usually based on all of the test results.

Note 2: This rule applies if both neoplasms are diagnosed simultaneously (at the same time) or less than 21 days apart.

Rule M12 Abstract as multiple primaries** when a neoplasm is **originally diagnosed** as a **chronic** (less aggressive) neoplasm **AND** there is a **second diagnosis** of an acute neoplasm **more than 21 days** after the chronic diagnosis.

Note 1: **This is a change from the pre-2010 rules**. Use the Heme DB Multiple Primaries Calculator to determine multiple primaries when a transformation from a chronic to an acute neoplasm occurs.

Note 2: Transformations are defined in the Heme DB

Example: Patient was diagnosed with MDS, unclassifiable in 2010. The patient presents in 2013 with a diagnosis of acute myeloid leukemia (AML). The transformation paragraph in the Heme DB says MDS (chronic neoplasm) transforms to AML (acute neoplasm). Because the diagnosis dates for the chronic neoplasm (MDS) and the acute neoplasm (AML) are more than 21 days apart, abstract two primaries, the MDS and the AML.

* Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

** Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

Rule M13 Abstract as multiple primaries[**] when both a **chronic** and an **acute** neoplasm are diagnosed **simultaneously or less than or equal to 21 days apart AND** there is **documentation of two** bone marrow examinations, lymph node biopsies, or tissue biopsies: **one confirming the chronic neoplasm and another confirming the acute neoplasm.**

Example: Vertebral biopsy on 2/13/2013 was positive for plasmacytoma and a 4/16/2013 bone marrow biopsy was positive for multiple myeloma. The biopsies and diagnoses were more than 21 days apart. Code as two primaries, solitary plasmacytoma of bone 9731/3 and plasma cell myeloma/multiple myeloma 9732/3.

Rule M14 Abstract the acute neoplasm as a single primary[*] when a neoplasm is **originally diagnosed** as **acute AND reverts** to a less aggressive/chronic neoplasm **AND** there is **no confirmation** available that the patient has been treated for the acute neoplasm.

Note 1: When these diagnoses happen **within 21 days**, it is most likely that the first diagnosis of acute neoplasm was a provisional diagnosis

Note 2: When the subsequent diagnosis occurs more than 21 days after the original diagnosis of acute neoplasm, it is important to follow-back to obtain information on treatment or a subsequent bone marrow biopsy that negates the diagnosis of acute neoplasm

Example: 3/16/2013 biopsy of cervical nodes was positive for diffuse large B-cell lymphoma (DLBCL) 9680. Bone marrow done on 4/18/2013 shows follicular lymphoma 9690. No treatment was given between the diagnoses of the chronic neoplasm (follicular lymphoma) and the acute neoplasm (DLBCL). Abstract one primary, DLBCL 9680.

Rule M15 Abstract multiple primaries[**] when a neoplasm is **originally diagnosed** as **acute AND reverts** to a less aggressive/**chronic** neoplasm **after treatment.**

Note 1: Only abstract as multiple primaries when the patient has been treated for the acute neoplasm

Note 2: **This is a change from the pre-2010 rules.** Use the Heme DB Multiple Primaries Calculator to determine multiple primaries when a transformation from an acute neoplasm to a chronic neoplasm occurs.

Note 3: Transformations are defined in the Heme DB.

Example: Patient was diagnosed in 2009 with AML. The patient was treated with chemotherapy and a subsequent stem cell transplant. After treatment, all diagnostic testing and bone marrow biopsies were negative for AML. On 2/25/2013 a bone marrow biopsy was positive for myelodysplastic syndrome. Abstract a second primary with the histology MDS 9989/3.

Rule M16 Use the Heme DB Multiple Primaries Calculator to determine the number of primaries for all cases that do **not** meet the criteria of M1-M15.

Example: Polycythemia vera (PV) diagnosed in 2001, receiving anagrelide. Increasing leukocytosis seen, bone marrow biopsy done in 2013 showing primary myelofibrosis (PMF) with myeloid metaplasia. The multiple primaries calculator shows that PV and PMF are two primaries.

[*] Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

[**] Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

Primary Site and Histology Coding Instructions and Rules

Primary Site and Histology Coding Instructions

1. Always use the Heme DB when coding primary site.
   - When a site code is listed under **Primary Site(s)**, that is the **only** primary site code that can be assigned for that leukemia, myelodysplastic syndrome, or myeloproliferative syndrome.
     Example: Acute myeloid leukemia 9861/3. Code the primary site to bone marrow C421.
   - For lymphoma, there will be rare exceptions to the site listed under **Primary Site(s)**. The following are some exceptions:
     - When lymphoma is present only in the bone marrow
     - When lymphoma is present only in a site not listed under **Primary Site(s)**
     - When a physician documents an organ or site of origin that is different than the one listed in the primary site paragraph
   - Read the Abstractors Notes when instructed to do so under **Primary Site(s)**. The Abstractor Notes contain the most common and also the rare primary sites for the histology.

2. Code **primary site** using:
   - Scans
   - Medical record documentation
   - Pathology report
   - Heme DB (as described in Step 1)
     Note: For hematopoietic neoplasms, the pathology report is not the default for determining the primary site. The standard for determining primary site differs depending upon the specific histology.

3. Code the **histology** from the **Definitive Diagnostic Method(s)** section of the Heme DB. Definitive diagnostic method(s) may be any of the following:
   - Clinical diagnosis
   - Genetic test
   - Immunophenotyping
   - Cytology
   - Pathology
     - Final diagnosis
     - Comment on final diagnosis
     - Addenda to final diagnosis
     - CAP protocol

4. When tests or reports defined as definitive diagnostic method(s) are **not available**, code the primary site and histology using the following documentation. The list is in **hierarchical order** starting with A.
   A. **Documentation** in the medical record referring to the **original** scans, genetic testing, immunophenotyping, or pathology reports
   B. **Documentation** in the medical record that refers to the histology and primary site
   C. Death certificate

5. Do **not** use ambiguous terms to code a specific histology.

Note 1: For hematopoietic and lymphoid neoplasms, the ambiguous terminology means a specific histology has not been determined
Note 2: Ambiguous terminology **is** used to determine reportability. The instructions for coding specific histologies and for reportability are different.
Note 3: Ambiguous terminology **IS** used for casefinding. It **cannot**, however, be used to assign a specific histology. The use of ambiguous terminology to identify a specific hematopoietic neoplasms usually indicates that the physician does not have enough proof to make a definitive diagnosis.
Example 1: Myeloproliferative disease, **probably** polycythemia vera; code MPN, NOS.
Example 2: CT scan, right neck swelling. Biopsy of mass was done and the final diagnosis on the pathology report is B-cell lymphoma, favor Burkitt lymphoma. Hemilaminectomy of spinal L2-L5 was done and the pathology report showed lymphoma, B-cell phenotype, favor Burkitt lymphoma. Code the histology to B-cell lymphoma, 9590/3. Do not use ambiguous terminology **to code histology** for hematopoietic neoplasms. "**Favor**" is ambiguous terminology.

6. For lymphomas only, use Module 7, Coding Primary Sites for Lymphomas.

Primary Site and Histology Coding Rules

Note 1: The primary site and histology coding rules are divided into 10 modules. Each **module** covers a group of **related** hematopoietic or lymphoid **neoplasms**. However, a specific histology may be covered in more than one module.

Note 2: Go to the first module that fits the case you are abstracting. If the situation in your case is not covered in that module, **continue** on **as directed** after the last rule in the module.

Note 3: The modules are **not hierarchical**, but the rules within each module are in **hierarchical** order. Apply the rules **within each module** in order. **Stop** at the first rule that applies.

Module 1: Post-Transplant Lymphoproliferative Disorder PH1 - PH2

Post-transplant lymphoproliferative disorder 9971/3

Rule PH1 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), or organ(s), and code the **histology post-transplant lymphoproliferative disorder 9971/3** when the diagnosis is **only** post-transplant lymphoproliferative disorder.

Note 1: These neoplasms are polymorphic post-transplant lymphoproliferative disorders. The diagnosis may or may not include the word polymorphic.
Note 2: The patient must have a history of a solid organ transplant or an allogenic bone marrow transplant.
Note 3: Most cases occur within a year of transplantation; however, they can occur anytime after the transplant.
Note 4: Polymorphic PTLD are caused by the post-transplant T-cell immunosuppressant drugs. Treatment for the polymorphic PTLD is the decrease or cessation of immunosuppressant drugs.

Rule PH2 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), or organ(s), and code the **histology of the accompanying lymphoma or plasmacytoma when the diagnosis is post-transplant lymphoproliferative disorder and any B-cell lymphoma, T-cell lymphoma, Hodgkin lymphoma, or plasmacytoma.**

Note 1: These neoplasms are monomorphic post-transplant lymphoproliferative disorders. The diagnosis may or may not include the word “monomorphic”.
Note 2: The patient must have a history of a solid organ transplant or an allogenic bone marrow transplant.
Note 3: Most cases occur within a year of transplantation; however, they can occur anytime after the transplant.
Note 4: Monomorphic PTLD is also caused by the immunosuppressant drugs. Patients are treated for the lymphoma or plasmacytoma.
Example: Bone marrow biopsy positive for B-cell lymphoma. Abdominal mass biopsy was positive for PTLD, monomorphic type and aggressive B-cell malignancy. Immunohistochemistry shows the B-cell malignancy to be Burkitt lymphoma. Code the histology to Burkitt lymphoma and primary site C42.1.

When this module does not apply to the case being abstracted, go to Module 7 .

Module 2: Plasma Cell Neoplasms PH3 – PH7

Extraosseous plasmacytoma 9734/3
Plasma cell leukemia 9732/3
Plasma cell myeloma/multiple myeloma 9732/3
Solitary plasmacytoma of bone 9731/3

Rule PH3 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), or organ(s), and code the **histology extramedullary plasmacytoma 9734/3** when any of the following occur in a site **other than bone:**

- Plasma cell neoplasm
- Extraosseous (extramedullary) plasmacytoma
- Multiple extraosseous (extramedullary) plasmacytomas
- Multiple plasmacytomas
- Plasmacytoma, NOS
- Solitary plasmacytoma

Note 1: Extramedullary and extraosseous mean "not occurring in bone"

Note 2: 80% of extramedullary plasmacytomas occur in the upper respiratory tract (oropharynx, nasopharynx, sinuses, and larynx) although they may occur in numerous other sites including the GI tract, lymph nodes, bladder, CNS, breast, thyroid, testis, parotid, and skin

Note 3: Do **not** code to blood C420, bone marrow C421, reticuloendothelial system, NOS C423, or the hematopoietic system, NOS C424

Example 1: Pathology reports a solitary plasmacytoma wrapped around L4 vertebrae, no invasion of vertebrae. Code the primary site as soft tissue C496 and the histology 9734/3.

Example 2: Scan shows two plasmacytomas in the nasopharyngeal wall. Biopsy confirms plasmacytoma. Code the primary site nasopharynx C119 and the histology 9734/3.

Rule PH4 Code the primary site to the **specific bone C400-C419** where the plasmacytoma originated and code the histology **solitary plasmacytoma** of bone **9731/3** when the diagnosis is:

- Multiple medullary plasmacytomas
- Multiple plasmacytomas
- Multiple plasmacytomas of bone
- Plasma cell neoplasm
- Solitary medullary plasmacytoma
- Solitary plasmacytoma
- Solitary plasmacytoma of bone

Note 1: The most common sites are bones with active bone marrow hematopoiesis; in order of frequency these include vertebrae, ribs, skull, pelvis, femur, clavicle, and scapula.

Note 2: Do **not** code primary site to blood C420, bone marrow C421, reticuloendothelial system, NOS C423, or the hematopoietic system, NOS C424.

Rule PH5 Code the primary site **bone, NOS C419** and histology **solitary plasmacytoma, NOS 9731/3** when the only information is that the patient had a **plasmacytoma, NOS** or a **solitary plasmacytoma**, NOS.

Note: When the only information available is that the patient had a plasmacytoma, default to coding plasmacytoma of bone.

Example: Death-certificate-only case with underlying cause of death listed as plasmacytoma.

Rule PH6 Code the primary site **bone marrow C421** and the histology plasma cell myeloma/**multiple myeloma 9732/3** when the **clinical** diagnosis is **plasma cell myeloma/multiple myeloma** and there is no documentation of bone marrow biopsy or the results of the bone marrow biopsy are unknown or unavailable

Note: A clinical diagnosis of multiple myeloma may be based on amyloidosis with associated renal impairment, anemia, and/or hypercalcemia supported by radiologic evidence of multiple lytic bone lesions.

Example: Death-certificate-only case with underlying cause of death listed as multiple myeloma.

Rule PH7 Code the primary **site bone marrow C421** and the histology **plasma cell myeloma/multiple myeloma 9732/3** when the diagnosis is:

- Evolving myeloma
- Indolent myeloma
- Multiple myeloma
- Plasma cell **leukemia**
- Plasma cell myeloma
- Smoldering myeloma

Note 1: The criterion for diagnosing multiple myeloma is equal to or greater than 10% of plasma cells in the bone marrow. This is an informative note only, the registrar does not code multiple myeloma based on bone marrow results. There must be a diagnosis of multiple myeloma.

Note 2: A medical record may have multiple bone marrow biopsies. If any one of the biopsies is positive for multiple myeloma, code the histology to multiple myeloma 9732/3 and the primary site to bone marrow C421.

Example: Bone marrow biopsies: Biopsy 1: Negative. Biopsy 2: Multiple myeloma with bone marrow showing 18% plasma cells. Code the primary site bone marrow C421 and the histology 9732/3.

Note 3: Plasma cell leukemia is now classified as a variant of multiple myeloma and coded 9732/3 multiple myeloma. It is irrelevant whether the plasma cell leukemia occurs before the multiple myeloma, simultaneously, or after the patient has been diagnosed with multiple myeloma.

When this module does not apply to the case being abstracted, go to Module 7.

Module 3: Lymphoma/Leukemia (Specific neoplasms that can manifest as either leukemia or lymphoma or both leukemia and lymphoma) PH8 – PH10

Adult T-cell leukemia/lymphoma (HTLV-1 positive) 9827/3
Adult T-cell leukemia/lymphoma 9837/3
B Lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL) 9816/3
B Lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3);E2A-PBX1 (TCF3-PBX1) 9818/3
B Lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1) 9814/3
B Lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH 9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1 9812/3
B lymphoblastic leukemia/lymphomas, NOS 9811/3, 9812/3-9818/3
Blastic natural killer leukemia/lymphoma 9727/3
Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL) 9823/3
Lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged 9813/3

Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma.
Note 2: Commonly lymphoma originates in lymph nodes, tissue, or an organ although it will metastasize to the bone marrow when the disease is stage IV or disseminated.
Note 3: Leukemia usually originates in the bone marrow.

Rule PH8 Code the **primary site** as indicated in the following bullets and code the histology **chronic lymphocytic leukemia/small lymphocytic lymphoma CLL/SLL 9823/3** when the diagnosis is **CLL, SLL,** or **CLL/SLL**.

- Code the **primary site** to **bone marrow C421** when the **bone marrow is involved** or when **only peripheral blood is involved**. (Lymph node(s) or lymph node region(s), organ(s), or tissue(s) may also be involved.)

OR

- Code the **primary site** to the **involved lymph node(s) or lymph node region(s), the involved organ(s) , or tissue(s)** when there is **no bone marrow involvement or when it is unknown if bone marrow is involved**.

Note 1: CLL will always have peripheral blood involvement. The bone marrow may or may not be involved. In later stages of the disease there may be involvement of lymph nodes, liver, and spleen.
Note 2: SLL will always have involvement of lymph node(s) or lymph node region(s), organ(s) or tissue(s). SLL was previously coded to 9670/3. Histology code 9670/3 is now obsolete because it is virtually impossible to distinguish SLL from CLL/SLL.
Note 3: CLL/SLL will have involvement of bone marrow AND involvement of lymph node(s) or lymph node region(s), organ(s), or tissue(s).
Note 4: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.
Note 5: See Appendix C for help identifying lymph node names, chains, regions and codes.
Note 6: See **Module 7** for help in coding primary site for lymphomas.

Rule PH9 Code the primary site **bone marrow C421** when the **only** site involved is **bone marrow**. Code the **histology using the list below.**

- Adult T-cell leukemia/lymphoma (HTLV-1 positive) 9827/3
- Adult T-cell leukemia/lymphoma 9837/3
- B Lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL) 9816/3

- B Lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3);E2A-PBX1 (TCF3-PBX1) 9818/3
- B Lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1) 9814/3
- B Lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH 9817/3
- B Lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1 9812/3
- B lymphoblastic leukemia/lymphoma, NOS 9811/3
- Blastic plasmacytoid dendritic cell neoplasm 9727/3
- Lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged 9813/3

Note: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH10 Code the primary site to the **site of origin,** lymph node(s) or lymph node region(s), tissue(s) or organ(s) involved. Code the **histology** using the list below.

- Adult T-cell leukemia/lymphoma (HTLV-1 positive) 9827/3
- Adult T-cell leukemia/lymphoma 9837/3
- B Lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL) 9816/3
- B Lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3);E2A-PBX1 (TCF3-PBX1) 9818/3
- B Lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1) 9814/3
- B Lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH 9817/3
- B Lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1 9812/3
- B lymphoblastic leukemia/lymphoma, NOS 9811/3
- Blastic plasmacytoid dendritic cell neoplasm 9727/3
- Lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged 9813/3

Note 1: Do **not** simply code the site of a biopsy; also use the information available from scans to determine the correct primary site. See Module 7 for more information on coding primary site for lymphomas.

Note 2: See Appendix C for help identifying lymph node names, chains, regions and codes.

When this module does not apply to the case being abstracted, go to Module 7.

Module 4: Preleukemia, Smoldering Leukemia, and Myelodysplastic Syndrome PH11

Preleukemia, smoldering leukemia, and myelodysplastic syndrome 9989/3

Rule PH11 Code the primary site **bone marrow C421** and the histology **myelodysplastic syndrome 9989/3** when the diagnosis is **preleukemia, smoldering leukemia, or myelodysplastic syndrome.**

When this module does not apply to the case being abstracted, go to Module 7.

Module 5: Myeloid Neoplasms and Mast Cell Neoplasms PH12 - PH15

Acute myeloid leukemia, NOS 9861/3, 9811/3-9897/3
Mast cell leukemia 9742/3
Mast cell sarcoma 9740/3
Myeloid sarcoma 9930/3

Rule PH12 Code the primary site **bone marrow C421** and code the **histology** to **mast cell leukemia 9742/3** when the diagnosis is **mast cell sarcoma AND** there is a simultaneous or previous diagnosis of **mast cell leukemia.**

Note: When mast cell sarcoma follows a diagnosis of mast cell leukemia, the sarcoma is a manifestation of late-stage leukemia. The mast cells infiltrate soft tissue.

Rule PH13 Code the primary site to the **site of origin** (usually soft tissue) and the **histology** to **mast cell sarcoma 9740/3** when the diagnosis is **mast cell sarcoma AND** there is **NO** previous or simultaneous diagnosis of **mast cell leukemia.**

Rule PH14 Code the primary site **bone marrow C421** and code the histology **acute myeloid leukemia, NOS or any of the specific AML histologies 9840/3, 9861/3, 9865/3-9967/3, 9869/3, 9891/3, 9895/3-9898/3, 9910/3, 9911/3 and 9931/3** when the diagnosis is **myeloid sarcoma AND** there is a simultaneous or previous diagnosis of acute myeloid leukemia.

Note 1: When myeloid sarcoma follows a diagnosis of acute myeloid leukemia the sarcoma is a manifestation of late-stage leukemia

Note 2: This is a change from the 2012 Hematopoietic manual. The following histologies were removed from this rule: 9820, 9823, 9826, 9827, 9831-9837, 9863.

Rule PH15 Code the primary site to the **site of origin** (usually soft tissue) and the histology to **myeloid sarcoma 9930/3** when the diagnosis is **myeloid sarcoma AND** there is **no** previous or simultaneous diagnosis of acute myeloid leukemia.

Note: This is a single primary coded to myeloid sarcoma. See Rule M3.

When this module does not apply to the case being abstracted, go to Module 7.

Module 6: Coding Primary Site for Specified Lymphomas PH16 - PH24

B-cell lymphoma, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma 9596/3
Diffuse large B-cell lymphoma 9680/3
Follicular lymphoma 9690/3
Follicular lymphoma, grade 1 9695/3
Follicular lymphoma, grade 2 9691/3
Follicular lymphoma, grade 3, 3A, 3B 9698/3
Lymphoplasmacytic lymphoma 9671/3
Primary cutaneous follicle center cell lymphoma 9597/3
Waldenstrom macroglobulinemia 9761/3

Rule PH16 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s) or organ(s) and code the histology **diffuse large B-cell lymphoma** (DLBCL) 9680/3 when **DLBCL** 9680/3 **AND follicular** lymphoma (NOS, grade 1, grade 2, or grade 3) are present in the same lymph node(s) or lymph node region(s), the same organ(s), the same tissue(s) or bone marrow. FL includes the following:

- FL, **NOS** 9690/3
- FL **grade 1** 9695/3
- FL **grade 2** 9691/3
- FL **grade 3** 9698/3

Note 1: Do **not** code the follicular lymphoma grade in the grade/differentiation field.

Note 2: The original pathology may identify only DLBCL although both DLBCL and follicular lymphoma are present. The DLBCL is much more aggressive than the follicular lymphoma and often masks the follicular lymphoma during the initial work-up. Because it is more aggressive, the DLBCL will respond more rapidly to treatment so the post-treatment biopsy may show a combination of DLBCL and follicular lymphoma or the post-treatment biopsy may be positive for only follicular lymphoma. The follicular lymphoma was present from the beginning but was hidden. Do not change the histology; it should remain 9680/3.

Note 3: Use this rule when the diagnosis of DLBCL and FL are **simultaneous OR** when the FL is diagnosed in a post-treatment biopsy. The timing for a post-treatment biopsy would be within or equal to 60 days after the completion of initial treatment.

Note 4: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Primary Site Coding Instructions and Module 7 for more information on coding primary site for lymphoma.

Note 5: See Appendix C for help identifying lymph node names, chains, regions and codes.

Rule PH17 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), or organ(s), and the histology to follicular lymphoma (see examples) when the diagnosis is diffuse follicular lymphoma or follicular lymphoma, diffuse.

Note: All variants of follicular lymphoma (NOS, grade 1, grade 2, and grade 3) were once called "follicle center lymphoma." Although that term is obsolete, it is sometimes used to describe follicular lymphoma. You will also see "follicle center" in the pathology reports for follicular lymphoma. However, the primary site and other sites of involvement will differ between follicular lymphoma and follicle center lymphoma. Follicle center lymphoma is a cutaneous malignancy with only rare involvement of regional lymph nodes. Follicular lymphoma commonly occurs in nodes and extranodal sites. (See the Heme DB Abstractor Notes for both neoplasms for information on clinical presentation and common primary sites.)

Example 1: Diffuse follicular lymphoma, grade 1. Code follicular lymphoma, grade 1 9695/3.

Example 2: Follicular lymphoma, diffuse, grade 2. Code follicular lymphoma grade 2 9691/3.

Example 3: Grade 3 follicular lymphoma, diffuse. Code follicular lymphoma, grade 3 9698/3.

Example 4: Follicular lymphoma, diffuse. Code follicular lymphoma, NOS 9690/3.

Rule PH18 Code the primary site to **skin** C44_ and the histology to **primary cutaneous follicle center cell lymphoma** 9597/3 when there is **skin infiltration** with **follicle cell lymphoma** or **B-cell lymphoma, follicle type** and the involvement is:

- Limited to **skin**
- Limited to **skin** and the **regional lymph node(s).**

Note 1: If there is involvement of lymph node(s) that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code follicle cell lymphoma and do **not** code skin as the primary site. Dissemination to other sites or distant lymph nodes is uncommon and would occur late in the stage of the disease.

Note 2: All variants of follicular lymphoma (NOS, grade 1, grade 2, and grade 3) were once called "follicle center lymphoma." Although that term is obsolete, it is sometimes used to describe follicular lymphoma. You will also see "follicle center" in the pathology reports for follicular lymphoma. However, the primary site and other sites of involvement will differ between follicular lymphoma and follicle center lymphoma. Follicle center lymphoma is a cutaneous malignancy with only rare involvement of regional lymph nodes. Follicular lymphoma commonly occurs in nodes and extranodal sites. (See the Heme DB Abstractor Notes for both neoplasms for information on clinical presentation and common primary sites.)

Rule PH19 Code the primary site to **skin** C44_ and the histology to **diffuse large B-cell lymphoma** 9680/3 when there is **skin infiltration** with **large B-cell lymphoma** or **B-cell lymphoma**, **large cell type** and the involvement is:

- Limited to **skin**
- Limited to **skin** and the **regional lymph node(s).**

Note: If there is involvement of lymph node(s) that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.

Rule PH20 Code the primary site to **skin** C44_ and the histology to **diffuse large B-cell lymphoma** 9680/3 when there is **skin infiltration** with **B-cell lymphoma, NOS** and the involvement is:

- Limited to **skin**
- Limited to **skin** and the **regional lymph node(s).**

Note: If there is involvement of lymph node(s) that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.

Rule PH21 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow and the histology B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma/**composite Hodgkin and non-Hodgkin lymphoma 9596/3** when **both** non-Hodgkin lymphoma and Hodgkin lymphoma are **simultaneously** present in the **same** lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow.

Note 1: Use the composite lymphoma code when

- Both NHL and HL are present in one lymph node or multiple lymph nodes in one lymph node region
- Both NHL and HL are present in multiple lymph nodes in one lymph node region or several lymph node regions as defined by ICD-O-3. i.e. NHL and HL present in superior hilum and superior rectal lymph nodes.
    - When only one node is biopsied, assume all lymph nodes are involved with both NHL and HL

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Primary Site Coding Instructions and Module 7 for more information on coding primary site for lymphoma.

Note 3: See Appendix C for help identifying lymph node names, chains, regions and codes.

Note 4: Do **not** use the composite lymphoma code 9596/3 when

- NHL is present in one node and HL in another node within the same chain i.e. NHL in one cervical lymph node and HL in another cervical lymph node.
- NHL is present in one lymph node region and HL is present in another lymph node region i.e. NHL in cervical lymph node(s) and HL in inguinal lymph node(s)
- NHL in liver and HL in intra-thoracic lymph nodes

Rule PH22 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow and the histology to the NHL with the **numerically highest ICD-O-3 code** when two or more **non-Hodgkin lymphomas** are present in the same lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow.

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Primary Site Coding Instructions and Module 7 for more information on coding primary site for lymphoma.

Note 2: See Appendix C for help identifying lymph node names, chains, regions, and codes

Note 3: This rule does **not** apply to an NOS and more specific histology

Note 4: This rule does **not** apply when different NHLs are present in different sites. Examples are

- Thymic extranodal marginal-zone B-cell lymphoma is present in the thymus and diffuse large B-cell lymphoma in the hilar lymph nodes.
- B-cell lymphoma is present in the intrathoracic lymph nodes and peripheral T-cell NHL in the liver.

Example: Biopsy performed on 9/9/2013 revealing both small lymphocytic lymphoma and follicular grade 2 lymphoma in the same LN. Per Rule M6, this is one primary. Code histology to 9823/3 since it is the numerically higher ICD-O-3 code.

Rule PH23 Code the primary site **blood** C420 and the histology **Waldenstrom macroglobulinemia** 9761/3 when there is:

- Clinical diagnosis of Waldenstrom macroglobulinemia **AND/OR**
- IgM monoclonal gammopathy in the blood **and/or** bone marrow

Note: There may be a mention of lymphoplasmacytic lymphoma (LPL) in the bone marrow biopsy or blood. LPL is an NOS code and Waldenstrom Macroglobulinemia is one of the two specific LPLs (Gamma heavy chain disease is the other).

Rule PH24 Code the primary site to the **involved bone marrow, lymph nodes, or tissue** and the histology **lymphoplasmacytic lymphoma** 9671/3 when:

- There is a clinical diagnosis of lymphoplasmacytic lymphoma **AND/OR**
- Flow cytometry on bone marrow, lymph node(s),or tissue is positive for IgG, IgA and IgM monoclonal gammopathy

When this module does not apply to the case being abstracted, go to Module 7.

Module 7: Coding Primary Site for Lymphomas Only PH25 - PH37

9590/3-9729/3, 9811/3-9818/3, 9823/3, 9827/3, 9837/3

Rule PH25 Code the primary site to the **specific lymph node region** when only **one lymph node or one lymph node region** is involved.

Rule PH26 Code the primary site to **mediastinal lymph nodes** C771 when the site of lymphoma is described **only** as a **mediastinal mass.**

Rule PH27 Code the primary site to **intra-abdominal lymph nodes** C772 when the site of lymphoma is described **only** as a **retroperitoneal mass** or **only** as a **mesenteric mass.**

Rule PH28 Code the primary site **to inguinal lymph nodes** C774 when the site of lymphoma is described **only** as an **inguinal mass.**

Rule PH29 Code the primary site to the **specific lymph node region** when **multiple** lymph node **chains** within the **same region** as defined by ICD-O-3 are involved.

Note: Use this rule when there is bilateral involvement of lymph nodes.

Example 1: Code intra-abdominal lymph nodes C772 when there is involvement of hepatic C772 and para-aortic lymph node chains C772.

Example 2: Code lymph nodes of head, face and neck C770 when there is involvement of cervical C770 and mandibular C770 lymph node chains.

Example 3: Code mediastinal lymph nodes C771 when bilateral mediastinal lymph nodes are involved.

Rule PH30 Code the **primary site** as **multiple lymph node regions, NOS** C778, when multiple lymph node regions, as defined by ICD-O-3, are involved and it is **not possible to identify the lymph node region where the lymphoma originated.**

Note 1: See Rule PH31 when there is also **organ** involvement.

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Primary Site Coding Instructions for more information on coding primary site for lymphoma.

Note 3: See Appendix C for help identifying lymph node names, chains, regions, and codes.

Example 1: Cervical C770 and intrathoracic C771 lymph nodes involved with B-cell lymphoma. Code the primary site to lymph nodes of multiple regions C778.

Example 2: CT scans showed involvement of the cervical lymph nodes C770 and the mediastinal lymph nodes C771. No additional involvement was identified during the work-up. Biopsy of a cervical lymph node confirmed lymphoma. Code the primary site to lymph nodes of multiple regions C778.

Rule PH31 Code the **primary site** to **lymph nodes, NOS** C779 when:

- Multiple lymph node region(s) **and organ(s)** are involved **AND**
- **No primary site** (lymph node region or organ) is identified.

Rule PH32 Code the **primary site** to **bone marrow** C421 when lymphoma is **present only in the bone marrow.**

Note: All available physical exams, scans, and other work-up must be negative for lymph node, tissue, or organ involvement OR no other workup was done

Example: Bone marrow biopsy is positive for diffuse B-cell lymphoma (DLBCL). No other work up performed. Code primary site to bone marrow. If further workup is done that identifies a primary site, reassign primary site.

Rule PH33 Code the **primary site** to the **organ** when lymphoma is present only in an **organ.**

Note 1: Includes lymphomas that are primary in the spleen. Splenic primaries are rare. Histologies that arise in the spleen include splenic marginal zone lymphoma 9689; hepatosplenic T-cell lymphoma 9716; splenic B-cell lymphoma/leukemia, unclassifiable 9591; splenic diffuse red pulp small B-cell lymphoma 9591; splenic marginal zone diffuse variant 9591; splenic EBV-associated B-cell lymphoproliferative disorder 9680. Follow-back for additional information when the histology is other than those listed **AND**

- The ***only information is a biopsy of the spleen OR***
- There is a physician statement that the spleen is the organ of origin

Note 2: Secondary involvement of the bone marrow would be coded in CS.

Example: Pathology from stomach resection shows lymphoma. No other sites of involvement are identified. Code the primary site to stomach, NOS C169.

Rule PH34 Code the **primary site t**o the **lymph node region** as defined by ICD-O-3 when there **is proof of extension from the regional lymph nodes** into an organ.

Example: Patient presents with abdominal adenopathy. Surgical exploration documents direct invasion of the stomach from the regional lymph nodes. Code abdominal lymph nodes C772.

Rule PH35 Code the primary site to the organ when a lymphoma is present in an organ and that organ's regional lymph nodes.

Note 1: In Stage II, III and IV disease, distant lymph nodes or other organs, such as spleen, may be involved. Disregard the distant lymph nodes and splenic involvement.
Note 2: Code the primary site to the organ. Use the Collaborative Stage Data Collections System to determine regional vs. distant lymph nodes.
Example 1: Lymphoma is present in the kidney and peri-renal lymph nodes. Code the primary site to kidney C649.
Example 2: Lymphoma is present in the stomach and the gastric lymph nodes. Code the primary site to stomach, NOS C169.
Example 3: Lymphoma is present in the spleen and the splenic lymph nodes. Code the primary site to spleen C422.

Rule PH36 Code **the primary site** to **lymph nodes, NOS** C779 when:

- Lymphoma is present in an organ and lymph nodes that are not regional for that organ and the origin cannot be determined even after consulting the physician **OR**
- Lymphoma is present in more than one organ and the regional nodes for all organs involved **OR**
- More than one organ and some combination of regional and distant nodes for the organs involved

Note 1: Lymphoma can spread from organs to **regional** lymph nodes, but does **not** spread from the organ directly to **distant** lymph nodes
Example: The patient has positive mediastinal C771 and cervical C770 lymph nodes and involvement of the stomach C169. No further information is available. Code to lymph node, NOS C779.
Note 2: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes
Note 3: See Appendix C for help identifying lymph node names, chains, regions, and codes

Rule PH37 Code **primary site** to **unknown** primary site C809 when there is no evidence of lymphoma in lymph nodes AND the physician **documents** in the medical record that he/she **suspects** that the lymphoma **originates** in an **organ(s) OR multiple organ involvement without any nodal involvement.** See ICD-O-3 Rule D.

When this module does not apply to the case being abstracted, go to Module 8.

Module 8: Multiple Histologies Coded as a Single Primary PH38 – PH39

Hodgkin and Non-Hodgkin Lymphoma AND Multiple non-Hodgkin Lymphomas

Rule PH38 Code **primary** site to involved lymph node(s) or lymph node region(s), organ(s), or tissue(s) and the **histology** to B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma/composite Hodgkin and non-Hodgkin lymphoma **9596/3** when any Hodgkin **AND** non-Hodgkin lymphoma are diagnosed simultaneously in the same site, lymph node(s) or lymph node region(s), organ(s), or tissue(s).

Rule PH39 Code the **primary** site to involved lymph node(s) or lymph node region(s), organ(s) or tissue(s) and the histology to the higher numeric ICD-O-3 code when two or more non-Hodgkin lymphomas are diagnosed simultaneously in the same site, lymph node(s) or lymph node region(s), organ(s), or tissue(s).
Note: Do **not** use this rule for an NOS and more specific lymphoma.

When this module does not apply to the case being abstracted, go to Module 9.

Module 9: NOS and More Specific Histology PH40 - PH41

All hematopoietic and lymphoid neoplasms 9590/3-9992/3

Rule PH40 Code the **non**-specific (**NOS**) histology when the diagnosis is:

- **One non**-specific histology **AND**
- **Two or more specific** histologies **AND**
- The Heme DB multiple primaries calculator documents the specific histologies and NOS are the **same primary AND**
- No further information is available

Note : Use the Heme DB multiple primaries calculator to confirm that the NOS and specific histologies are the same primary

Example 1: The diagnosis is myelodysplastic/myeloproliferative neoplasm unclassifiable 9975/3, polycythemia vera 9950/3, essential thrombocythemia 9962/3. The Heme DB multiple primaries calculator shows that myelodysplastic/myeloproliferative neoplasm unclassifiable and polycythemia vera are the same primary. The multiple primaries calculator also shows that myelodysplastic/myeloproliferative neoplasm unclassifiable and essential thrombocythemia are the same primary. Follow-back produces no additional information. Code the histology myeloproliferative disorder, NOS 9960/3.

Example 2: Pathology report states morphologic features and immunophenotype of low grade B-cell lymphoma are most compatible with lymphoplasmacytic lymphoma or marginal zone lymphoma. The term "compatible with LBL 9671/3 or MZL 9699/3" means that the immunophenotype was not diagnostic for either disease. Default to the NOS, the B-cell lymphoma, 9591.

Rule PH41 Code the **specific** histology when the diagnosis is:

- **One non**-specific (NOS) histology **AND**
- **One specific** histology **AND**
- The Heme DB multiple primaries calculator documents the specific histology and NOS are the **same primary**

Note : Use the Heme DB multiple primaries calculator to confirm that the NOS and specific histology are the same primary

When this module does not apply to the case being abstracted, go to Module 10.

Module 10: Coding Primary Site and Histology PH42 - PH43

All hematopoietic and lymphoid neoplasms 9590/3-9992/3

Use Only When Modules 1-9 are Not Applicable

Rule PH42 Use the **Heme DB** to determine the primary site and histology when rules PH1-PH41 do **not** apply.

Note: For primary site, use the information under Primary Site(s) in the Heme DB **and/or** the abstractor notes as instructed in the introduction to this Manual

Rule PH43 Code the histology to **the numerically higher** ICD-O-3 code when the histology code cannot be determined using the Heme DB.

Note: This rule should rarely be used

This is the end of the rules for coding primary site and histology.

Grade of Tumor Rules

Priority List for Coding Grade or Phenotype

This is a hierarchical list with Note 1 having the highest priority.

Note 1: Use the Grade of Tumor Rules (G1-G9) to assign the correct code in the grade field.
If the pathology report states a different grade than the ones stated for G1-G4, use the rules from the Hematopoietic manual.
Note 2: Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade. This table is outdated.
Note 3: Use a physician's statement to code the phenotype in the grade field, use statements from **any part** of medical record including but not limited to:

- Pathology report
- History and physical
- Consultation
- Final diagnosis
- Face sheet

Note 4: When there is no physician statement, code Grade/Phenotype 9 Unknown.
Note 5: The **only** valid grade codes for hematopoietic neoplasms are 5, 6, 7, 8, **AND** 9.
Note 6: Do **not** code descriptions "low grade," "intermediate grade," or "high grade" in the Tumor Grade field. These terms refer to the Working Formulation categories of lymphoma diagnosis.

Rule G1 Code cell type not determined, not stated, not applicable, **code 9**, for the following myeloproliferative neoplasms, myeloproliferative/myelodysplastic syndromes, myelodysplastic syndrome, histiocytic and dendritic cell neoplasms:

9740/3	9945/3
9741/3	9946/3
9742/3	9950/3
9751/3	9961/3
9755/3	9962/3
9756/3	9963/3
9757/3	9964/3
9758/3	9975/3
9759/3	9980/3
9801/3	9982/3
9805/3	9983/3
9806/3	9985/3
9807/3	9986/3
9808/3	9989/3
9809/3	9991/3
9875/3	9992/3
9876/3	

Note 1: These neoplasms do not have a specific codable phenotype
Note 2: See Tables B1, B3, B4, and B11 in Appendix B for neoplasm terms and codes

Rule G2 Code T-cell, **code 5,** for the following neoplasms; **T-cell** is part of the neoplasm name or the neoplasm is of **T-cell origin**

9700/3
9701/3
9702/3
9705/3
9708/3
9709/3
9714/3 (unless pathologist specifically designates as a B-cell)
9716/3
9717/3
9718/3
9724/3
9725/3
9726/3
9827/3
9834/3
9837/3

Note 1: Record T-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention T-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.

Note 2: When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Rule G3 Code B-cell, **code 6**, for the following **B-cell precursor lymphoid neoplasms and the mature B-cell neoplasms:**

9591/3	9687/3	9731/3	9815/3
9596/3	9688/3	9732/3	9816/3
9597/3	9689/3	9734/3	9817/3
9670/3	9690/3	9737/3	9818/3
9671/3	9691/3	9738/3	9823/3
9673/3	9695/3	9762/3	9826/3
9678/3	9698/3	9811/3	9833/3
9679/3	9699/3	9812/3	9836/3
9680/3	9712/3	9813/3	9940/3
9684/3	9728/3	9814/3	

Note 1: Record B-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention B-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.

Note 2: When the medical record or pathology report contains one of these terms with a different phenotype (T-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Note 3: See Tables B7 and B8 in Appendix B.

Rule G4 Code **NK-cell** (natural killer cell), **code 8**, for the following neoplasms:

9719/3
9948/3

Note 1: Reco**rd NK-**cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention NK-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.

Note 2: When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, T-cell, or null-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Note 3: See Table B9 in Appendix B.

Rule G5 Code T-cell, **code 5**, when the neoplasm is identified as **T-cell, T-cell phenotype, T-precursor, Pre-T, gamma-delta-T, or null-cell and T-cell.**

Rule G6 Code B-cell, **code 6**, when the neoplasm is identified as **B-cell, B-cell phenotype, B-precursor, pre-B, or null-cell and B-cell.**

Rule G7 Code Null cell, non-T non-B, **code 7**, when the neoplasm is described as **null cell, non-T non-B,** or **common cell.**

Rule G8 Code Natural Killer (NK) cell, **code 8**, when the neoplasm is described as **NK cell, natural killer cell, nasal NK/T-cell lymphoma, or null-cell and NK cell.**

Rule G9 Code cell type not determined, not stated, not applicable, **code 9**, when:

- There is **no statement describing the cell type OR**
- The cell type is described as **combined T AND B cell OR**
- The cell type is described as **combined B AND NK cell**

Glossary

Acute neoplasm: For the purposes of these rules, the term “acute neoplasm” means that another neoplasm , a chronic neoplasm, transform to this neoplasm.

Allogeneic bone marrow/stem cell transplant: Marrow is used from a donor whose human leukocyte antigens (HLA) closely match the patient’s. Stem cells are taken either by bone marrow harvest or by aphaeresis from a genetically matched donor. (See also Bone marrow transplant and Stem cell transplant.)

Anemia: The number of red blood cells is below normal for blood count.

Anemia of chronic disorder/disease: NOT REPORTABLE. This form of anemia is caused by patients with chronic diseases such as arthritis or chronic infections. It is also caused by malignancies. The anemia itself is not a malignancy. This form of anemia accounts for approximately 25% of all anemias.

Apheresis: Process in which the blood of a donor or patient is passed through an apparatus that separates out one particular constituent and returns the remainder to circulation.

Autologous bone marrow/stem cell transplant: The patient’s own bone marrow is used. (See also Bone marrow transplant and Stem cell transplant.)

B-cells: B-cells make antibodies against antigens and are produced in the bone marrow and mature in the bone marrow. B cells are lymphocytes that play a large role in the humoral immune response (as opposed to the cell-mediated immune response, which is governed by T-cells).

B-cell and T-cell (combined): Because a major loss or dysfunction of T cells can cause secondary B-cell deficiency, a number of disorders show clinical manifestations of combined B- and T-cell deficiency, though the only pathology is in the T-cell. In contrast, some diseases appear to primarily involve the T-cells and do not appear to affect antibody production. Those diseases are discussed in T-cell disorders.

B-cell leukemia: A type of cancer that forms in B-cells. This neoplasm usually occurs in adults and may be indolent (slow growing) or aggressive (fast growing). Many different types of B-cell lymphomas and leukemias have been identified. (See also B-cells.)

B-cell lymphoma: A type of cancer that forms in B-cells. B-cell lymphomas usually occur in adults and may be either indolent (slow-growing) or aggressive (fast-growing). There are many different types of B-cell lymphomas, and prognosis and treatment depend on the type and stage of cancer. (See also B-cells.)

BCR-ABL cancer gene: A mutant gene that is formed when a piece of chromosome 9 attaches to the end of chromosome 22. The BCR-ABL cancer gene gives the cell instructions to make a protein that leads to CML.

Bence Jones protein: A protein made by myeloma cells that is found in the plasma and urine of many patients with myeloma. This type of protein is also called "light chains" because it represents a smaller segment of the whole immunoglobulin molecule, composed of heavy and light chains.

Biopsy: The removal of a sample of tissue for purposes of diagnosis. (Many definitions of "biopsy" stipulate that the sample of tissue is removed for examination under a microscope. This may or may not be the case. The diagnosis may be achieved by other means such as by analysis of chromosomes or genes.)

Blast cell: Immature blood-forming cell.

Blood chemistry tests: These measure the amount of certain chemicals in the blood. They are not used to diagnose leukemia; rather, they detect liver or kidney damage caused by leukemic cells or the chemotherapy drugs.

Blood count (CBC or complete blood count): A count of the number of red blood cells, white blood cells and/or platelets in a given sample of blood. (See also Peripheral blood smear).

Bone marrow aspiration: The removal of a small amount of bone marrow (usually from the hip) through a needle. The needle is placed through the top layer of bone and a liquid sample containing bone marrow cells is drawn into a syringe. The sample is then sent for cytologic examination. Bone marrow aspiration is done to diagnose and follow the progress of various conditions, including anemia and cancer, and to obtain marrow for transplantation.

Bone marrow biopsy: The removal of a sample of bone marrow and a small amount of bone (usually from the hip) through a large needle. The sample is a core biopsy to obtain bone marrow together with bone fibers. The sample is examined under a microscope (pathology) to see the cells and architecture of the bone marrow.

Bone marrow transplant: A complex treatment that may be used when cancer is advanced or has recurred. The bone marrow transplant makes it possible to use very high doses of chemotherapy that would otherwise be impossible because they destroy the patient's normal marrow cells as well. When used for advanced or recurrent cancer, a portion of the patient's or a donor's bone marrow is withdrawn, purged of tumor cells if possible (in the case of autologous transplantation), and stored. Then the patient is given high doses of chemotherapy to kill the cancer cells. But the drugs also destroy the remaining bone marrow, thus robbing the body of its natural ability to fight infection. The stored marrow is given by transfusion (transplanted) to rescue the patient's immune defenses. Currently most patients are treated with stem cell transplants rather than bone marrow transplants.

Bone marrow: The soft tissue in the internal portion of flat bones and vertebrae that produces new blood cells.

CBC: (See Blood count.)

CD; **cluster of differentiation**: Used to define the findings in immunophenotyping (See also Immunophenotyping). The CD is the antigen found in the cell and is associated with a number such as CD16 which may be present on leukemic lymphoblasts.

Chromosome: A part of the cell that carries genes. Genes give instructions that tell the cell what to do. The cell has 46 chromosomes, 22 pairs of chromosomes plus two sex chromosomes. Females have two "X" chromosomes. Males have one "X" and one "Y" chromosome.

Chromosome abnormality or mutation: Chromosomal anomalies may reflect an atypical number of chromosomes (see also karyotyping) or a structural abnormality in one or more chromosomes. Chromosome anomalies usually occur when there is an error in cell division following mitosis or meiosis. Many types of chromosome anomalies exist, including single chromosome mutations such as deletion, duplication and inversion. The two major chromosome mutations include: insertion (see also chromosome insertion) or translocation (see also chromosome translocation) of genes.

Chromosome deletion: A portion of the chromosome is missing or deleted.

Chromosome duplication: A portion of the chromosome is duplicated, resulting in extra genetic material.

Chromosome insertion: Insertions can vary in size from one base pair incorrectly inserted into a DNA sequence to a large section of one chromosome inserted into another chromosome. Basically, it is the loss of a portion of one chromosome with the "lost" portion inserted into a different chromosome.

Chromosome inversion: A portion of the chromosome has broken off, turned upside down, and reattached; therefore the genetic material is inverted. In contrast to chromosome insertion where a different chromosome is inserted into another chromosome, chromosome inversion uses the same chromosome. Basically it is a switch of a small portion of one chromosome with a small portion of another chromosome so all of the genetic material is retained, but the two small portions of chromosomes have changed places with each other.

Chromosome translocation: Translocation occurs when a portion of one chromosome is transferred to another chromosome. There are two main types of translocations; reciprocal and Robertsonian. In a reciprocal translocation, segments from two different chromosomes have been exchanged. In a Robertsonian translocation, an entire chromosome has attached to another chromosome at the centromere.

Chromosome: A part of the cell that carries genes. Genes give instructions that tell the cell what to do. The cell has 46 chromosomes, 22 pairs of chromosomes plus two sex chromosomes. Females have two "X" chromosomes. Males have one "X" and one "Y" chromosome.

Chronic neoplasm: For the purposes of these rules, the term “chronic neoplasm” means a neoplasm that transforms to another, more acute neoplasm.

Clinical diagnosis: A diagnosis made on the basis of physician expertise; there is no microscopic or imaging confirmation. For hematopoietic and lymphatic neoplasms, clinical diagnosis includes diagnoses of exclusion where microscopic and imaging confirmation is equivocal and the physician uses his/her expertise to evaluate the clinical presentation and equivocal tests to make a diagnosis.

Colony-stimulating factors (CSF): Types of growth factors that promote growth and division of blood-producing cells in the bone marrow. CSFs are naturally produced in the body. Note: CSF agents are classified as immunosuppressing drugs, but are not coded on the cancer abstract.

Combined modality therapy: Two or more types of treatment used alternately or together to improve results. For example, surgery for cancer is often followed by chemotherapy to destroy any cancer cells that may have spread from the original site, or local radiation is given to treat any cancer cells that may have been left behind after surgery.

Complete remission: A term that is applied to a patient's health status after treatment, when there is no sign of the disease using standard tests specific for that disease and the patient has returned to good health.

Consolidation therapy: A term usually applied to the treatment of acute leukemia for drug treatment given to patients in remission after induction therapy. The aim of consolidation therapy is to kill as many of the remaining cancer cells as possible.

Cutaneous lymphoma: Lymphoma originating in the skin.

Cytochemistry: After cells are placed on slides they are exposed to chemical stains (dyes) that are attracted to or react with some types of leukemia cells.

Cytogenetic analysis: The term for a lab test that is used to examine the chromosomes in marrow, blood, and lymph node cells. It can confirm that the cells are cancer cells (malignant) and in some cases the results may guide the intensity of therapy.

Cytogenetic response (cytogenetic remission): A treatment response in which there are no leukemia, lymphoma or myeloma cells detected in the blood and/or marrow by the FISH test.

Cytogenetics: The study of chromosomes, the visible carriers of DNA. This is a fusion science joining cytology, the study of cells, with genetics, the study of inherited variation.

Cytokines: Products of cells of the immune system that regulate the immunologic, inflammatory and reparative responses. Some may stimulate immunity and cause the regression of cancers.

Cytostatic: Describes the way some anti-cancer drugs work, not what type of drug they are. Cytostatic treatments stop the cancer cells from multiplying; they do not kill cancer cells.

Cytotoxic: Toxic to cells; cell-killing. Cytotoxic describes any agent or process that kills cells. Chemotherapy and radiotherapy are forms of cytotoxic therapy.

Definitive diagnosis: For the purpose of these rules, the definitive diagnosis is the diagnostic method or methods listed in the Heme DB (under the category "definitive diagnostic method"). The definitive diagnoses are the methodologies used to identify the specific neoplasm.

Dendritic cell: The dendrites are immune cell with threadlike tentacles which enmesh antigen; T cells then attack the antigen. Both Langerhans cells, found in the skin and follicular dendritic cells, found in lymphoid tissues, and are types of dendritic cells.

Differentiation: The normal process through which cells mature so they can carry out the jobs they were meant to do. Cancer cells are generally less differentiated than their normal counterparts. Differentiation also refers to the greater or lesser degree of morphologic similarity that cancer cells have to normal cells.

DNA: Abbreviation for deoxyribonucleic acid. DNA holds genetic information on cell growth, division, and function.

Donor lymphocyte infusion: A treatment that uses an infusion of white cells called lymphocytes from the original stem cell donor.

Drug resistance: When a drug used to treat a patient's disease does not work or stops working.

Electrophoresis, serum, protein, fluorescent and urine: A type of test that separates substances, especially proteins, and analyzes molecular structure. Electrophoresis may be used to provide a confirmation of a diagnosis of multiple myeloma. Electrophoresis is used to detect M proteins in the urine or serum (M-spike).

Epstein Barr Virus (EBV): Discovered by Epstein, Achong, and Barr by electron microscopy from Burkitt lymphoma tissue. EBV is also called Human herpes virus 4 (HHV-4). EBV is a precursor to several kinds of cancer.

Etiology: The cause of a disease. Cancer has many causes, although research is showing that both genetics and lifestyle are major factors in many cancers.

Extramedullary myeloma: A tumor of plasma cells in a site other than bone marrow.

Extramedullary: Occurring outside any of the medullas including the medulla oblongata and the medullary cavities of the bones.

Extraosseous: Occurring outside of the bone or bones.

FISH: Analysis can be done on blood, bone marrow, lymph nodes, and any other tissue. The short name for a test called "fluorescence in situ hybridization," a test to measure the presence in cells of a specific chromosome or gene. This test can be used to plan treatment and to measure the results of treatment.

Flow cytometry: A method of counting types of cells with fluorescent tags on the surfaces of the cells. Flow cytometry is often used to determine the type of leukemia, lymphoma or myeloma cells that are present. For example, each disease subtype has a specific pattern of markers on its cell surface. Flow cytometry can also detect residual levels of disease after treatment.

Gene: Parts of cells that give instructions for making proteins. Proteins help the cell do its job.

Genetic testing: Analysis can be done on blood, bone marrow, lymph nodes, and any other tissue. Laboratory studies of blood or other tissue to analyze DNA in order to identify chromosome abnormalities which identify specific neoplasms. Genetic testing is also done to Identify genetic alternation that may indicate an increased risk for developing a specific disease or disorder.

Genotype: The genetic makeup, as distinguished from the physical appearance, of an organism or a group of organisms.

Hematologic response (hematologic remission): A treatment response where the leukemia, lymphoma or myeloma cell numbers are decreased in the blood; and, red cell count, white cell count, and platelet count are either at or near normal values.

Hematologist: A physician who treats blood cell diseases.

Hematopoietic: Pertaining to hematopoiesis which means the production of all types of blood cells.

Hematopoietic Neoplasms (as used in these rules): Includes leukemias, lymphomas, and mast cell tumors (9590-9992).

Hemolytic anemia: NOT REPORTABLE. A group of acute or chronic anemias, inherited or acquired, characterized by shortened survival of mature erythrocytes and inability of bone marrow to compensate for the decreased life span.

Hodgkin lymphoma (HD), Hodgkin disease (HD), Hodgkin's: A cancer of the lymphatic system. Hodgkin lymphoma appears to originate in a lymph node and later spreads to the spleen, liver, and bone marrow. The presence of the Reed-Sternberg cell distinguishes Hodgkin lymphoma from non-Hodgkin lymphoma.

Human leukocyte antigens (HLA): HLA tissue typing identifies the best tissue/blood cell match between donors and recipients.

ICD-O: The International Classification of Diseases for Oncology published by the World Health Organization (WHO). This reference is used to code the primary site and histology for reportable neoplasms in a standardized manner. These codes allow historic and current data to be grouped for reporting and analysis.

Immune response: The reaction of the body to foreign material such as an infection-causing microorganism, an immunization, or the cells of another individual used for an allogeneic stem cell transplant.

Immune system: The complex system by which the body resists infection by microbes such as bacteria or viruses. The immune system may also help the body fight some cancers. It is also responsible for the rejection of transplanted tissues or organs.

Immunocytochemistry: Cells from the bone marrow are treated with special antibodies that cause certain types of cells to change color. Immunocytochemistry is sometimes helpful in determining the exact type of acute leukemia. It is not necessary in most cases of chronic leukemia.

Immunophenotyping: Analysis can be done on blood, bone marrow, lymph nodes, and any other tissue. A sample of blood, bone marrow cells, or lymph node cells is analyzed to determine the types of antigens or markers on the surface of the cell. This analysis is used to diagnose specific types of leukemia and lymphoma, for example, myelogenous leukemic cells can be distinguished from lymphomatous leukemic cells. The antigen in the cell is usually referred to as CD followed by a number (See definition of CD).

Immunosuppression: A state in which the body's immune system does not respond as it should. This condition may be present at birth, or it may be caused by infections [such as human immunodeficiency virus (HIV)] or by cancer therapies such as cancer-cell killing (cytotoxic) drugs, radiation, and bone marrow transplantation.

Induction therapy: The initial treatment of a patient with a blood cancer with chemotherapy (or radiation therapy). The aim of induction therapy is to kill a maximum number of blood cancer cells so as to induce a remission (absence of signs or effects of the disease).

In situ lymphoma: NOT REPORTABLE In situ lymphoma is a recent diagnosis and the criteria for diagnosing an in situ leukemia have not been established and accepted by the hematologic community. We will defer collection until those criteria are established and accepted.

JAK2, Janus kinase 2, Exon 12: JAK2 is a gene mutation that increases susceptibility to several myeloproliferative neoplasms (MPNs). Testing for the JAK2 mutation is done on whole blood or on bone marrow. Nearly all people with polycythemia vera and about half of those with essential thrombocythemia and primary myelofibrosis have the mutation. When JAK2 is positive, the MPN is definitely reportable; however JAK2 does not identify which specific MPN is present. Correlation for other clinical findings and laboratory findings is needed to determine the specific histology. The JAK2 mutation has also been found in a minority of patients with other myeloid stem cell disorders, including chronic myelomonocytic leukemia.

Karyotyping: To arrange and classify chromosomes based on number, size, shape, and other characteristics.

Late effects: Medical problems that do not develop or become apparent until months or years after treatment ends. Examples of late effects include the development of a treatment-related cancer or heart disease.

Leukapheresis: A process in which extra white cells are removed by a machine.

Leukemia: Cancer of the blood or blood-forming organs. People with leukemia often have a noticeable increase in white blood cells (leukocytes) in the peripheral blood.

Leukocyte alkaline phosphatase (LAP) test: Useful in distinguishing CML from other types of leukemia and benign conditions.

Leukopenia: Decrease in the while blood cell count, often a side effect of chemotherapy.

Light chains: A part of the monoclonal (M) protein in myeloma. Monoclonal immunoglobulin (protein), like normal immunoglobulin is usually made up of two heavy (larger) chains and two light (smaller) chains attached to each other. The abnormal production of immunoglobulin protein by myeloma cells sometimes results in parts of the molecule that is the heavy chain or light chain, being made and discharged from the myeloma cells. They can each be measured in plasma. The light chains are small enough to pass through the kidney and enter the urine, where they can be detected.

Long-term effects: Medical problems that persist for months or years after treatment ends, for example, infertility, growth problems in children, or cancer treatment-related fatigue.

Lymph nodes (lymph glands): Small bean-shaped collections of immune system tissue such as lymphocytes and macrophages found along lymphatic vessels. The nodes remove cells and cell waste from lymph and help fight infections.

Lymph: Clear fluid that flows through the lymphatic vessels and contains cells known as lymphocytes. These cells are important in fighting infections and may also have a role in fighting cancer.

Lymphatic organ: An organ of the immune system where lymphocytes develop and congregate such as tonsils, adenoids, thymus and spleen. (There is disagreement among the experts on the division of lymphatic organs and lymphatic tissues. For the purpose of these rules, we use them as synonyms meaning any aggregate of lymph cells within the body).

Lymphatic system: The tissues and organs, including the bone marrow, spleen, thymus, and lymph nodes, that produce and store cells that fight infection and disease.

Lymphatic tissue: Immune system tissue where lymphocytes develop and congregate such as tonsils, adenoids, thymus and spleen. (There is disagreement among the experts on the division of lymphatic organs and lymphatic tissues. For the purpose of these rules, we use them as synonyms meaning any aggregate of lymph cells within the body).

Lymphatic vessels: These channels connect the lymph nodes. They contain lymph - a fluid that carries lymphocytes as they circulate from one lymph node area to another. The lymphatic channels are connected to the blood vascular system permitting lymphocytes to enter the blood.

Lymphoblastic: A term used to describe a type of blood cell disease caused by young or immature lymphocytes or "lymphoblasts". An example is acute lymphoblastic leukemia, which is characterized by the presence of malignant (cancerous) lymphoblasts (immature lymphocytes).

Lymphocyte: A type of white blood cell that is part of the immune system. There are three major types of lymphocytes: B lymphocytes that produce antibodies to help combat bacterial, fungal or viral infections; T lymphocytes that have several functions, including assisting B lymphocytes to make antibodies; and natural killer (NK) cells that can attack virus-infected cells or tumor cells.

Lymphocytic: A term used to describe a type of blood cell abnormality caused by lymphocytes. An example is chronic lymphocytic leukemia, which is characterized by the presence of malignant (cancerous) lymphocytes. Sometimes used as a synonym for "lymphoblastic."

Lymphoma: A type of cancer that begins with a malignant change in a lymphocyte, lymph node cell or a cell in the lymphatic tissue of the marrow, gastrointestinal tract, spleen, skin or other sites.

M protein (monoclonal protein): Myeloma cells make a protein called monoclonal immunoglobulin, sometimes referred to as M protein. M protein, like normal immunoglobulin is usually made up of two heavy (larger) chains and two light (smaller) chains attached to each other. The amount of M protein in the blood can be measured in the laboratory. It is used to estimate the extent of the myeloma and to follow the effects of treatment.

Marrow: The spongy center inside of bones.

Matched donor: A person whose major tissue types are identical to those of a patient who is seeking a stem cell transplant. The patient can be given the donor's healthy matched stem cells, which can restore blood and immune cells after high-intensity cancer treatment.

Molecular genetic studies: The branch of genetics that deals with hereditary transmission and variation on the molecular level.

Molecular response: A treatment response is called a complete molecular remission if no leukemia cells in the blood and/or marrow can be detected by PCR.

Monoclonal antibody therapy: A type of therapy that targets and kills cancer cells. Monoclonal antibodies are immune proteins made in the laboratory. They are designed to target to a specific blood cancer cell. They produce less toxic effects on normal tissues than chemotherapy.

Monocyte: A type of white cell. Monocytes and neutrophils are the two major microbe-eating and killing cells in the blood.

Multiple myeloma (MM), myeloma, Kahler disease, myelomatosis, plasma cell myeloma: A type of cancer that begins in plasma cells (white blood cells that produce antibodies). MM is characterized by excessive numbers of abnormal plasma cells in the bone marrow and overproduction of intact monoclonal immunoglobin.

Myelofibrosis: A progressive disease of the bone marrow. Neoplastic bone marrow stem cells lodge and grow in multiple sites outside the bone marrow. Symptoms include enlargement of the spleen and a gradual replacement of the bone marrow elements by fibrosis (scarring), progressive anemia, and variable changes in the number of white blood cells and platelets.

Myelogenous: A term used to describe a form of blood cancer that begins in a marrow stem cell or early marrow progenitor cell. A blood cancer that begins in the marrow is called leukemia. Myelogenous leukemias usually do not directly affect lymphocytes. The terms "myeloid" or "myelocytic" are sometimes used instead of "myelogenous."

Myeloma: A tumor of antibody-producing cells, called plasma cells, which are normally found in the bone marrow.

Myelodysplasia: MAY BE REPORTABLE. Dysplasia of myelocytes. This term may be used as a synonym for myelodysplastic syndrome, however, the terms MDS, or myelodysplastic syndrome should be used somewhere in the medical record.

Natural killer cell (NK cell): NK cells, like killer T cells, attack and kill cancer cells and cells infected by microorganisms. NK cells can react against and destroy another cell without prior sensitization to that cell. Natural killer (NK) cells are part of our first line of defense against cancer cells and virus-infected cells. NK cells are small lymphocytes that originate in the bone marrow.

Neutropenia: A decrease below normal in the concentration of neutrophils, a type of white cell.

Neutrophil: A type of white cell. Neutrophils and monocytes are the two main microbe-eating cell and infection-fighting in the blood.

Non-Hodgkin lymphoma (non-Hodgkins, NHL): A cancer of the lymphatic system. What distinguishes non-Hodgkin lymphoma from Hodgkin lymphoma (Hodgkin disease) is the absence of a type of cell called the Reed-Sternberg cell. This cell is present only in Hodgkin lymphoma.

Non-myeloablative stem cell transplant (mini-transplant): A type of allogeneic stem cell transplant that does not use high-dose chemotherapy as a treatment. The patient takes special drugs so that his or her immune system does not reject the transplanted stem cells. Over a long time, the donated cells replace the patient's

blood and immune system cells. The donated cells also attack the leukemia, lymphoma or myeloma cells. Other drugs to help the transplanted stem cells fight the blood cancer without attacking healthy cells are being tested in clinical trials.

Nucleus: A part of the cell containing the chromosomes and genes.

Null cell: A lymphocyte that develops in the bone marrow and lacks the characteristic surface markers of the B and T lymphocytes. A null cell is a large granular lymphocyte without surface markers or membrane-associated proteins from B lymphocytes or T lymphocytes. Natural killer cells are usually null cells with surface marker CD 16, which binds to the Fc portion of the IgG, thereby destroying it.

Oncologist: A physician who treats patients with cancer.

Patch mycosis fungoides: The early stage of mycosis fungoides is called the "patch stage." The skin is infiltrated by patches or lumps of lymphocytes (white cells). In this early stage, the neoplasm appears in ovoid patches that do not cover large amounts of skin. During the patch stage of mycosis fungoides it is difficult to distinguish the neoplasm from eczema or psoriasis.

Pathologist: A doctor who examines cells and tissues obtained from biopsies to determine the type of disease present.

PCR: The short name for a lab test called "polymerase chain reaction," a very sensitive test that can measure the presence of a blood cancer cell marker in the blood. It is used to detect remaining blood cancer cells that are below the detection of cytogenetic methods (e.g. FISH).

Peripheral blood smear: Blood is viewed under a microscope to count different circulating blood components (red cells, white cells, platelets, etc.) to see whether the cell population level is normal. (See also Blood count.)

Phenotype: The observable traits or characteristics of an organism, for example hair color, weight, or the presence or absence of a disease. Phenotypic traits are not necessarily genetic.

Plasma: Part of the blood that is mostly water, with some vitamins, minerals, proteins, hormones and other natural chemicals.

Plasmacytoma, extraosseous/extramedullary: Localized plasma cell neoplasms that arise in tissue other than bone.

Plasmacytoma, solitary plasmacytoma of bone/osseous plasmacytoma: Localized bone tumor consisting of monoclonal plasma cells.

Platelet (thrombocyte): A type of blood cell that prevents bleeding and forms a plug that stops bleeding after an injury to the body.

Ploidy: Degree of repetition of the base number of chromosomes in a cell. A diploid cell has double the base number, one set from each parent, while a monoploid cell has only one set (a haploid is a sex cell, which also has only one set). Aneuploidy is an abnormal condition in which a chromosome is missing or an extra one is inserted.

Polymerase chain reaction (PCR): PCR is a molecular technique which allows the production f large quantities of a specific DNA from a DNA template.PCR has transformed the way that most studies requiring the manipulation of DNA fragments and DNA cloning may be performed.

Port: An implanted port is a type of long-term catheter. The port is surgically inserted under the skin's surface on the upper chest wall. After the site heals, no dressings are needed and no special home care is required. When medicines are needed, a physician, physician assistant or nurse inserts a needle through the skin to access the port. The patient can choose to have a local numbing cream applied to the injection site before the port is used. Blood can be drawn, and blood products can be received through this device.

Primary cutaneous B-cell lymphoma (PCBCL): B-cell lymphoma originating in the skin. There is no evidence of extracutaneous involvement at the time of diagnosis or for six months after diagnosis. Two subtypes of PCBCL have been identified by the European Organization for Research and Treatment of Cancer: follicular center cell lymphoma and large B-cell lymphoma.

Protocol: A formal outline or plan, such as a description of what treatments a patient will receive and exactly when each should be given and in what doses.

Radiation therapy: The use of x-rays or other high-energy rays to kill cancer cells.

Radioimmunotherapy: A treatment that uses antibodies to carry a radioactive substance to cancer cells to kill them. They are used in the treatment of lymphoma and lymphocytic leukemia.

Red cell: A blood cell that carries oxygen and delivers it to the body.

Reed Sternberg cells: Giant cells found on light microscopy in biopsies from patients with Hodgkin lymphoma and certain other disorders. They are derived from B lymphocytes. They are named after Dorothy Reed Mendenhall and Carl Sternberg who provided the first definitive microscopic descriptions of Hodgkin lymphoma.

Refractory disease: Disease that does not respond to therapy.

Relapse or recurrence: When disease comes back after it has been successfully treated.

Remission: A period of time with no signs of disease and/or when the patient does not have any symptoms of the disease.

Richter's transformation, Richter's syndrome (RS): Transformation of B-cell chronic lymphocytic leukemia (CLL) or hairy cell leukemia (HCL) to diffuse large B-cell lymphoma.

Risk factor: A factor that may increase the chance that a person will develop a disease or condition. For example, cigarette smoking is a risk factor for lung cancer.

Side effect: The signs or symptoms a patient may have from the effects of treatment on healthy cells.

Signs and symptoms: A sign is a change in the body that the doctor sees in an exam or a lab test. A symptom is a change in the body that a patient can see or feel.

Skin infiltration: For lymphoma diagnoses, skin infiltration means involvement of the skin (not spread to the skin). The term "skin infiltration" is commonly used with cutaneous lymphomas. Cutaneous lymphomas originate in the skin.

Smoldering leukemia, preleukemia, myelodysplastic syndrome: The myelodysplastic syndromes are a group of diseases in which the bone marrow does not make enough healthy blood cells.

S-phase fraction (SPF): The percentage of cells that are replicating their DNA. DNA replication usually means that a cell is getting ready to divide into two new cells. In a tumor, a low SPF is a sign that a tumor is slow-growing; a high SPF shows that the cells are dividing rapidly and the tumor is growing quickly.

Spleen: The spleen is a lymphatic organ which is encased in a thick capsule. Within the spleen are two types of tissue: the white pulp, which is a lymphoid tissue and usually surrounds the blood vessels, and the red pulp, which is a network of channels (sinuses) that are filled with blood. The white pulp has typical lymphoid elements such as plasma cells, lymphocytes, and lymphatic nodules, which help fight infection. The red pulp destroys old red blood cells and filters the blood.

Stem cell: A type of cell found in marrow that makes red cells, white cells and platelets.

Stem cell transplant: A variation of bone marrow transplantation in which immature blood cells called stem cells are taken from the blood of the patient or a donor. Later, in the lab, the cells are stimulated with growth factors to produce more stem cells that are then transfused to the patient. Most hematopoietic diseases are treated with stem cell transplants rather than a bone marrow transplant.

Syngeneic bone marrow transplant: Transplant in which an identical twin is the bone marrow donor.

Systemic therapy: Treatment that reaches and affects cells throughout the body; for example, chemotherapy.

T cells: One type of white blood cell that attacks virus-infected cells, foreign cells, and cancer cells. T cells also produce a number of substances that regulate the immune response. T cells are produced in the bone marrow and mature in the thymus. They are the most common type of lymphocyte, itself divided into at least three subpopulations on the basis of function: cytotoxic, or killer, T lymphocytes; helper T lymphocytes; and suppressor T-lymphocytes.

T-cell lymphoma: A disease in which lymphoid cells called T-cells (or T lymphocytes) becomes malignant. T-cell lymphomas account for a minority (about 15%) of non-Hodgkin lymphomas in the US. The T-cell lymphomas are highly diverse and include lymphoblastic lymphoma (mainly in children and adolescents), peripheral T-cell lymphoma (a heterogeneous group of generally aggressive diseases), mycosis fungoides (called Sezary syndrome if the malignant T cells circulate in blood), and anaplastic large cell lymphoma (ALCL), which can be primary cutaneous ALCL or systemic ALCL. T-cell lymphomas account for about half of pediatric lymphomas.

Thrombocyte: Another word for platelet.

Thrombocytopenia: A decrease below normal in the number of platelets.

Vaccine therapy: A type of treatment under study for leukemia, lymphoma, or myeloma. This type of vaccine would not prevent the disease. The vaccine would increase the immune system's attack against cancer cells that remain after treatment with drugs.

Waldenstrom macroglobulinemia: A clinical diagnosis, not a pathology diagnosis. Most cases are associated with lymphoplasmacytic lymphoma, but occasionally it is associated with other subtypes of NHL. This is the only hematopoietic disease coded to blood (C420).

Watch and wait: An approach in which a physician closely observes a patient's condition with periodic medical exams and lab tests, without giving drugs or other forms of treatment for the disease in question. For some patients with non-growing or very slow-growing disease and no symptoms, watch and wait may be

preferred; it allows the patient to avoid drug treatment and its potential side effects until drugs are needed. This approach is based on studies that indicate early treatment in the specific situation in question is not beneficial.

White cell: Type of cell that fights infections in the body. There are two major types of white cells: germ-ingesting cells (neutrophils and monocytes) and lymphocytes, which provide an immune response to infection.

World Health Organization (WHO): The directing and coordinating authority for health within the United Nations. It is responsible for providing leadership on global health matters, shaping the health research agenda, setting norms and standards, articulating evidence-based policy options, providing technical support to countries, and monitoring and assessing health trends (from http://www.who.int/about/en/).

Appendix A
History of Hematopoietic and Lymphoid Neoplasm Coding

History of Coding Lymphoid Tissue and Hematopoietic System Neoplasms

Historically, diseases of lymphoid tissues and the hematopoietic system were believed to be separate entities, and the coding structure of the International Classification of Diseases was developed with this in mind. Prior to the early 1990s, the classification systems for lymphomas described malignant cells by their morphologic characteristics; for example, the size and shape of the tumor cell and its pattern of tumor growth and spread. The *International Classification of Diseases for Oncology,* Third Edition (ICD-O-3) says this about the historic classifications:

> Over the past 50 years many classifications of leukemia and lymphoma have been proposed. Some of these had a major impact on clinical practice while others are now largely forgotten. For most of this period, however, the distinction between lymphoma and leukemia has been regarded as of fundamental importance and classifications have tended to evolve separately (ICD-O-3, p. 13).

The World Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues, 4th Edition, was published in 2008. The 4th Edition of this world-renowned reference describes the current standard classification system for tumors of the hematopoietic and lymphoid systems. The 2008 classification continues to be based on the principles originally outlined in the REAL classification system (grouping by phenotype). These principles have now been applied to the classification of myeloid, lymphoid, mast cell, and histiocytic/dendritic neoplasms. Additionally, when specialized testing demonstrates one or more disease-specific or disease-defining characteristic using immunophenotyping and/or genetic testing, these characteristics have been incorporated into the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues,* 4th Edition classification system. Occasionally, a diagnosis may be based primarily on characteristic histologic features alone or in combination with clinical characteristics of the disease such as the presence or absence of a virus. Therefore, any combination of disease-specific characteristics may be described microscopically (histology/morphology), or may be identified by immunohistocytochemistry test, or identified by a specific immunophenotype or genetic abnormality. Part or all of these descriptive characteristics may be included in a new or updated hematopoietic or lymphoid neoplasm term or description (preferred term or synonym) or even in the disease classification (group) to which a specific disease entity may be assigned.

Several newly recognized conditions have been added to the 2008 classification. In addition, some conditions previously classified as borderline malignancy are now to be treated as malignant disease. The current classification divides hematopoietic and lymphoid neoplasms according to lineage. Three primary lines are used in the classification: myeloid, lymphoid, and histiocytic/dendritic. The 2008 *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues,* 4th Edition is used as the basis for this coding manual. The coding manual includes tables that describe the classification of disease along cell lines (lineage tables). Lineage tables are included in Appendix D.

The World *Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues,* 3rd Edition, published in 2001, was based on principles defined in the Revised European-American Classification of Lymphoid Neoplasms (REAL), originally published by the International Lymphoma Study Group in 1994. Both the REAL and current classifications group borderline and malignant tumors into broad categories by hematologic lineage: myeloid, lymphoid, histiocytic/dendritic, and mast cell. Within these broad categories or phenotypes, tumors may present in solid or circulating phases. Solid phase is the presence of malignant cells in tissue, such as lymph nodes, soft tissues, or organs; generally these have historically been called lymphomas. The circulating phase is characterized by the presence of malignant cells in the circulating blood or bone marrow; historically these have been called leukemias. According to the introduction to the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues,* 4th Edition, the "…distinction between them (lymphomas and

leukemias) is artificial. Thus B-cell chronic lymphocytic leukemia and B-cell small lymphocytic lymphoma are simply different manifestations of the same neoplasm, as are lymphoblastic lymphomas and lymphoblastic leukemias and Burkitt lymphoma and Burkitt leukemia" (2001 WHO Classification, page 13).

Although each of these pairs of diagnoses is histopathologically the same malignant cell with different presentations, they have different morphology code numbers in the *International Classification of Diseases for Oncology*, Third Edition (ICD-O-3). This is because ICD-O-3 is a subset of the *International Statistical Classification of Diseases and Related Health Problems*, Tenth Revision (ICD-10), in which the distinction between lymphomas and leukemias was maintained. ICD-10 was originally published in 1990, prior to the publication of the REAL classification that introduced the concept of grouping lymphoid and hematopoietic malignancies by phenotype rather than morphologic characteristics and clinical presentation. In order to ensure compatibility with ICD-10, the Third Edition of ICD-O differs from the structure of the WHO classification of hematologic malignancies.

The concept of cross-referencing two histology codes in ICD-O-3 was necessary because ICD-10 had not yet caught up with current medical concepts in the area of classification of lymphoma and leukemia. The following is noted in the introductory text of ICD-O-3 (page 14):

> ***Compatibility with ICD-10***
> In order to ensure compatibility with ICD-10, there are a number of ways in which the Third Edition of ICD-O differs from the structure of the WHO classification of hematologic malignancies. Separate codes have been allocated to B-cell chronic lymphocytic leukemia and B-cell small lymphocytic lymphoma. These are now recognized to be exactly the same entity, and for presentation of data these categories may therefore be combined. The same argument applies to lymphoblastic lymphoma and acute lymphoblastic leukemia, which are now regarded as the same disease but for which separate codes are provided.

The existence of dual codes for the same WHO classification entities is further discussed in the first errata for ICD-O-3 (5-22-2001):

> ***6. Assigning topography for hematopoietic diseases*** According to the medical understanding on which the World Health Organization Classification of Hematopoietic Neoplasms is based, some lymphomas and leukemias are the same disease with different presentations. For example, the WHO Classification lists B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (BCCLL/SLL) as a single entity, the same disease at different stages. The hemato-pathologists on the ICD-O-3 development committee recommended a single code number to represent the disease. However, since ICD-O is a subset of ICD-10 and ICD-10 is used to code mortality throughout the world, if a single ICD-O-3 code were used, there would be no way to determine whether a death was due to lymphoma or leukemia which are coded separately in ICD-10. As a result, it was necessary to retain separate codes for chronic lymphocytic leukemia and small lymphocytic lymphoma and link them. Thus, for the first time in ICD-O editions, some single disease entities are listed in two different categories and cross-referenced with the notation (see also M-9----). The topographic or primary site code for a diagnosis such as BCCLL/SLL depends on where the disease is diagnosed: if disease is diagnosed only in the blood or bone marrow, code the primary site to C42.1, bone marrow and assign the leukemia morphology code. For purposes of analysis according to the WHO Classification, cases from both morphology codes should be aggregated.

Resources used

World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues. 4th Edition, World Health Organization, 2008.
World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues. 3rd Edition, World Health Organization, 2001.
International Classification of Diseases for Oncology, Third Edition. World Health Organization, 2000.
Essential Haematology, Fifth edition. Hoffbrand AV, Moss PAH and Pettit JE. Blackwell Publishing, 2006.
Abstracting and Coding Guide for the Hematopoietic Diseases. National Cancer Institute, 2002.

Obsolete Terms as Defined in ICD-O-3
Hematopoietic and Lymphoid Neoplasms

The following tables identify terms that are no longer used to describe diseases and to display the current term used for that disease. The terms designated obsolete [obs] are based on ICD-O-3 term and category assignment only. Obsolete [obs] designations have not been updated to match the 2008 WHO Classification. Revised obsolete [obs] term designations matching the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition will be available when a revision or new edition of ICD-O is published either as an addendum to ICD-O-3 or as ICD-O-4. Note that the ICD-O-3 code does not change, only the name that is commonly used to describe the disease. The tables also provide information on the origin of the term and the date the term became obsolete.

( ) indicates an optional term in the phrase

Table A1: Histiocytic and Dendritic Cell Neoplasm Obsolete Terms

Obsolete Term	Notes	ICD-O-3 Code	Current Term
Histiocytic medullary reticulosis	*Term used as early as 1939; Obsolete as of 1987 with publication of Langerhans cell histiocytosis terminology*	9750/3	Malignant histiocytosis
Nonlipid reticuloendotheliosis	*Term used as early as 1955; Obsolete as of 1987 with publication of Langerhans cell histiocytosis terminology*	9754/3	Langerhans cell histiocytosis, disseminated

Table A2: Hodgkin Lymphoma (Hodgkin Disease) Obsolete Terms

Obsolete Term	Notes	ICD-O-3 Code	Current Term
Hodgkin disease, lymphocytic predominance, diffuse	*Source: Lukes-Butler classification, 1966; Obsolete: REAL classification 1994*	9651/3	Lymphocyte-rich classical Hodgkin lymphoma
Hodgkin disease, lymphocytic predominance, NOS	*Source : Rye classification, 1966 ; Obsolete : REAL classification 1994*	9651/3	Lymphocyte-rich classical Hodgkin lymphoma
Hodgkin disease, lymphocytic-histiocytic predominance	*Source: Lukes-Butler classification, 1966; Obsolete as of REAL classification 1994*	9651/3	Lymphocyte-rich classical Hodgkin lymphoma
Hodgkin paragranuloma	*Source : Jackson-Parker Classification, 1944 ; Obsolete : 1966*	9659/3	Nodular lymphocyte predominant Hodgkin lymphoma
Hodgkin paragranuloma, NOS	*Source : Jackson-Parker Classification, 1944 ; Obsolete : 1966*	9659/3	Nodular lymphocyte predominant Hodgkin lymphoma

Table A3: Lymphoid Neoplasm Obsolete Terms

Classification	Obsolete Term	Current Term	ICD-O-3 Code
B-cell Neoplasms	Immunocytoma	Lymphoplasmacytic lymphoma	9671/3
	Plasmacytoid lymphoma		
	Plasmacytic lymphoma		
	Mantle zone lymphoma	Mantle cell lymphoma	9673/3
	Intermediate differentiation diffuse lymphocytic lymphoma		
	Centrocytic lymphoma		
	Histiocytic lymphoma, NOS	Diffuse large B-cell lymphoma	9680/3
	Large cell cleaved and noncleaved lymphoma		
	Large cell diffuse lymphoma, NOS		
	Large cleaved cell lymphoma, NOS		
	Large cell cleaved lymphoma, NOS		
	Noncleaved diffuse lymphoma, NOS		
	Burkitt tumor	Burkitt lymphoma	9687/3
	Undifferentiated lymphoma, Burkitt type		
	Small noncleaved lymphoma, Burkitt type		
	Acute leukemia, Burkitt type	Burkitt cell leukemia	9826/3
	B-ALL		
	FAB L3		
	Centroblastic-centrocytic follicular lymphoma	Follicular lymphoma	9690/3
	Nodular lymphoma, NOS		
	Nodular lymphocytic lymphoma, NOS		
	Mixed small cleaved and large cell follicular lymphoma	Follicular lymphoma, grade 2	9691/3
	Mixed lymphocytic-histiocytic nodular lymphoma		
	Mixed cell type follicular lymphoma		
	Mixed cell type nodular lymphoma		
	Small cleaved cell follicular lymphoma	Follicular lymphoma, grade 1	9695/3
	Lymphocytic poorly differentiated nodular lymphoma		
	Large cell noncleaved follicular lymphoma	Follicular lymphoma, grade 3A Follicular lymphoma, grade 3B	9698/3
	Histiocytic nodular lymphoma		
	Noncleaved cell follicular lymphoma, NOS		
	Large cleaved cell follicular lymphoma		
	Lymphocytic well differentiated nodular lymphoma		
T-Cell and NK-Cell Neoplasms	Angiocentric T-cell lymphoma	Extranodal NK/T cell lymphoma, nasal type	9719/3
	Malignant reticulosis, NOS		

Classification	Obsolete Term	Current Term	ICD-O-3 Code
	Malignant midline reticulosis		
	Polymorphic reticulosis		
	Large cell (Ki-1 positive) lymphoma	Anaplastic large cell lymphoma, ALK positive	9714/3
	Peripheral T-cell lymphoma, AILD (Angioimmunoblastic Lymphadenopathy with Dysproteinemia)	Angioimmunoblastic T-cell lymphoma	9705/3
	Angioimmunoblastic lymphoma		

Table A4: Myeloid Neoplasm Obsolete Terms

Classification	Obsolete Term	Notes	Current Term	ICD-O-3 Code
Acute Myeloid Leukemias	Acute erythemia	*Listed as separate code in ICD-O-1; code changed to 9840 in ICD-O-3; Obsolete: as of FAB classification 1986*	Acute myeloid leukemia, M6 type	
	Di Guglielmo disease	*Eponym from as early as 1928; listed as synonym for acute erythemia in ICD-O-1 9840/3*	Acute erythroid leukemia	9840/3
	Acute erythremic myelosis	*Listed as synonym for acute erythemia in ICD-O-1*		
	Malignant myelosclerosis	*Term first used in 1963; Obsolete: as of FAB classification 1986*	Acute panmyelosis with myelofibrosis	9931/3
Chronic Myeloproliferative Diseases	Chronic erythemia	*Term used as early as 1892; not in ICD-O-1 or ICD-O-2; obsolete: 2001*	Polycythemia vera	9950/3
Myelodysplastic/ Myeloproliferative Diseases	Chronic myelomonocytic leukemia in transformation	*Source: French American British classification 1986; Obsolete: 2001*	Chronic myelomonocytic leukemia	9945/3
Myelodysplastic Syndromes	Preleukemia	*Term used as early as 1949; not in ICD-O-1; listed as synonym of MDS in ICD-O-2*	Myelodysplastic syndrome, unclassifiable	9989/3
	Preleukemic syndrome	*Term first used in 1973; not in ICD-O-1; listed as synonym of MDS in ICD-O-2*		

REFERENCES
1956 Publication of Rappaport classification of non-Hodgkin lymphomas
1966 Publication of Rye classification of Hodgkin lymphomas
1982 Publication of Working Formulation
1986 Publication of revised FAB classification (variously reported as 1982, 1985, or 1986)
2001 Publication of WHO classification, 3rd ed., and implementation of ICD-O-3
UICC TNM Supplement, 3rd ed., Wittekind, Greene, Henson, Hutter, Sobin

Appendix B
WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues
Histology Lineage

Use the Hematopoietic Database to identify synonyms that correspond to the WHO Preferred Term.

Table B1: Myeloproliferative Neoplasms

WHO Preferred Term	ICD-O-3
Chronic eosinophilic leukemia, NOS	9964/3
Chronic myelogenous leukemia, BCR-ABL1 positive	9875/3
Chronic neutrophilic leukemia	9963/3
Essential thrombocythemia	9962/3
Mast cell leukemia	9742/3
Mast cell sarcoma	9740/3
Myelodysplastic/myeloproliferative neoplasm unclassifiable	9975/3
Polycythemia vera	9950/3
Primary myelofibrosis	9961/3
Solitary mastocytoma of skin	9740/1
Systemic mastocytosis	9741/3

Table B2: Myeloid and Lymphoid Neoplasms with Eosinophilia and Abnormalities of PDGFRA, PDGFRB or FGFR1

WHO Preferred Term	ICD-O-3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Myeloid and lymphoid neoplasm with PDGFRA rearrangement	9965/3
Myeloid neoplasm with PDGFRB rearrangement	9966/3

Table B3: Myelodysplastic / Myeloproliferative Neoplasms

WHO Preferred Term	ICD-O-3
Atypical chronic myeloid leukemia, BCR-ABL1 negative	9876/3
Chronic myelomonocytic leukemia	9945/3
Juvenile myelomonocytic leukemia	9946/3
Myelodysplastic/myeloproliferative neoplasm, unclassifiable	9975/3
Refractory anemia with ring sideroblasts	9982/3

Table B4: Myelodysplastic Syndromes

WHO Preferred Term	ICD-O-3
Myelodysplastic syndrome associated with isolated del(5q)	9986/3
Myelodysplastic syndrome, unclassifiable	9989/3
Refractory anemia	9980/3
Refractory anemia with excess blasts	9983/3
Refractory anemia with ring sideroblasts	9982/3
Refractory cytopenia with multilineage dysplasia	9985/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3

Table B5: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms

WHO Preferred Term	ICD-O-3
Acute myeloid leukemias with recurrent genetic abnormalities	
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFB-MYH11	9871/3
Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26;2); RPN1-EVI1	9869/3
Acute myeloid leukemia with t(8;21)t(q22;q22); RUNX1-RUNX1T1	9896/3
Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL	9897/3
Acute promyelocytic leukemia (AML with t(15;17)(q22;q12), PML/RARA	9866/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3
Acute myeloid leukemia with myelodysplasia-related changes	9895/3
Therapy-related myeloid neoplasm	9920/3
Acute myeloid leukemia, NOS	9861/3
Acute monoblastic and monocytic leukemia	9891/3
Acute myeloid leukemia with minimal differentiation	9872/3
Acute myeloid leukemia without maturation	9873/3
Acute myeloblastic leukemia with maturation	9874/3
Acute myelomonocytic leukemia	9867/3
Acute erythroid leukemia	9840/3
Acute megakaryoblastic leukemia	9910/3
Acute basophilic leukemia	9870/3
Acute panmyelosis with myelofibrosis	9931/3
Myeloid sarcoma	9930/3

WHO Preferred Term	ICD-O-3
Myeloid proliferations related to Down syndrome	*No Code*
Transient abnormal myelopoiesis	9898/1
Myeloid leukemia associated with Down syndrome	9898/3
Blastic plasmacytoid dendritic cell neoplasm	9727/3

Table B6: Acute Leukemia of Ambiguous Lineage

WHO Preferred Term	ICD-O-3
Acute undifferentiated leukemia	9801/3
Mixed phenotype acute leukemia with t(v;11q2); MLL rearranged	9807/3
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Natural killer (NK) cell lymphoblastic leukemia/lymphoma	*No Code*

Table B7: Precursor Lymphoid Neoplasms

WHO Preferred Term	ICD-O-3
Adult T-cell leukemia/lymphoma	9837/3
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with recurrent genetic abnormalities	No Code
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A-PBX1 (TCF3-PBX1)	9818/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3

Table B8: Mature B-Cell Neoplasms

WHO Preferred Term	ICD-O-3
ALK positive large B-cell lymphoma	9737/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma	9596/3
B-cell prolymphocytic leukemia	9833/3
Burkitt lymphoma	9687/3
Chronic lymphocytic leukemia/small lymphocytic lymphoma	9823/3
Diffuse large B-cell lymphoma (DLBCL)	9680/3
Extranodal marginal zone lymphoma of mucosal-associated lymphoid tissue (MALT	9699/3

WHO Preferred Term	ICD-O-3
lymphoma)	
Extraosseous plasmacytoma	9734/3
Follicular lymphoma	9690/3
Hairy cell leukemia	9940/3
Heavy chain disease	9762/3
Intravascular large B-cell lymphoma	9712/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Lymphomatoid lesion	9766/1
Lymphoplasmacytic lymphoma	9671/3
Mantle cell lymphoma	9673/3
Non-Hodgkin lymphoma, NOS; splenic B-cell lymphoma/leukemia, unclassifiable	9591/3
Plasma cell myeloma	9732/3
Plasmablastic lymphoma	9735/3
Primary cutaneous follicle center lymphoma	9597/3
Primary effusion lymphoma	9678/3
Primary mediastinal (thymic) large B-cell lymphoma	9679/3
Solitary plasmacytoma of bone	9731/3
Splenic marginal zone lymphoma	9689/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3
Waldenstrom Macroglobulinemia	9761/3

Table B9: Mature T-Cell and NK-Cell Neoplasms

WHO Preferred Term	ICD-O-3
Adult T-cell leukemia/lymphoma (HTLV-1 positive)	9827/3
Aggressive NK-cell leukemia	9948/3
Anaplastic large cell lymphoma, ALK positive	9714/3
Angioimmunoblastic T-cell lymphoma	9705/3
Enteropathy-associated T-cell lymphoma	9717/3
Extranodal NK-/T-cell lymphoma, nasal type	9719/3
Hepatosplenic T-cell lymphoma	9716/3
Hydroa vacciniforme-like lymphoma	9725/3
Lymphomatoid papulosis	9718/1
Mycosis fungoides	9700/3
Peripheral T-cell lymphoma, NOS	9702/3
Primary cutaneous CD30 positive T-cell lymphoproliferative disorders	9718/3
Primary cutaneous T-cell lymphoma	9709/3
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Sezary syndrome	9701/3

WHO Preferred Term	ICD-O-3
Subcutaneous panniculitis-like T-cell lymphoma	9708/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
T-cell large granular lymphocytic leukemia	9831/3
T-cell prolymphocytic leukemia	9834/3

Table B10: Hodgkin Lymphoma

WHO Preferred Term	ICD-O-3
Classical Hodgkin lymphoma	9650/3
Lymphocyte-depleted classical Hodgkin lymphoma	9653/3
Lymphocyte-rich classical Hodgkin lymphoma	9651/3
Mixed cellularity classical Hodgkin lymphoma	9652/3
Nodular lymphocyte predominant Hodgkin lymphoma	9659/3
Nodular sclerosis classical Hodgkin lymphoma	9663/3

Table B11: Histiocytic and Dendritic Cell Neoplasms

WHO Preferred Term	ICD-O-3
Disseminated juvenile xanthogranuloma	No Code
Fibroblastic reticular cell tumor	9759/3
Follicular dendritic cell sarcoma	9758/3
Histiocytic sarcoma	9755/3
Interdigitating dendritic cell tumor	9757/3
Langerhans cell histiocytosis	9751/3
Langerhans cell sarcoma	9756/3

Table B12: Post-Transplant Lymphoproliferative Disorders (PTLD)

WHO Preferred Term	ICD-O-3
Early lesions	*No Code*
Classical Hodgkin lymphoma type PTLD	*
Monomorphic PTLD (B- and T/NK-cell types)	*
Plasmacytic hyperplasia	9971/1
Post-transplant lymphoproliferative disorder	9971/3

*These lesions are classified according to the leukemia or lymphoma to which they correspond, and are assigned the respective ICD-O code.

Appendix C
Lymph Node/Lymph Node Chain Reference Table

Use this table with the Primary Site and Histology Rules to determine whether involved lymph nodes are in a single ICD-O-3 lymph node region or in multiple ICD-O-3 lymph node regions.

This table contains the names of lymph nodes that have the capsule and sinus structure of true lymph nodes. Lymphoid tissue such as that in the GI tract, tonsils, etc., is not represented in this table.

Note: Pathology reports may identify lymph nodes within most organs, the most common being breast, parotid gland, lung, and pancreas. The lymph nodes in these organs are called intra-(organ name) lymph nodes such as intramammary lymph nodes. We have included the most common intra-organ lymph nodes on this table. For an intra-organ lymph node not listed on the table, code to the ICD-O-3 topography code for that organ's regional lymph node chain(s).

Table C1: Lymph Node/Lymph Node Chain Reference Table

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Abdominal	C772	Intra-abdominal	Mesenteric
Anorectal	C775	Pelvic	Pelvic, right and left*
Anterior axillary	C773	Axilla or arm	Axillary, right and left*
Anterior cecal	C772	Intra-abdominal	Mesenteric
Anterior deep cervical	C770	Head, face and neck	Cervical, right and left*
Anterior jugular	C770	Head, face and neck	Cervical, right and left*
Aortic NOS; ascending aortic lateral aortic; lumbar aortic; para-aortic; peri-aortic	C772	Intra-abdominal	Para-aortic
Aortico-pulmonary window (subaortic)	C772	Intra-abdominal	Para-aortic
Appendiceal	C772	Intra-abdominal	Mesenteric
Ascending aortic	C772	Intra-abdominal	Para-aortic
Aselli's glands (nodes near pancreas)	C772	Intra-abdominal	Para-aortic
Auricular NOS; infra-auricular; pre-auricular; post-auricular; retro-auricular	C770	Head, face and neck	Cervical, right and left*
Axillary, lateral;	C773	Axilla or arm	Axillary, right and left*
Axillary; anterior	C773	Axilla or arm	Axillary, right and left*
Azygos (lower paratracheal)	C771	Intrathoracic	Mediastinal
Brachial	C773	Axilla or arm	Axillary, right and left*
Bronchial; bronchopulmonary; hilar; proximal lobar; pulmonary root	C771	Intrathoracic	Hilar
Bronchopulmonary	C771	Intrathoracic	Hilar
Bronchopulmonary; bronchial hilar; proximal lobar; pulmonary root	C771	Intrathoracic	Hilar
Buccal	C770	Head, face and neck	Cervical, right and left*
Buccinator (facial)	C770	Head, face and neck	Cervical, right and left*
Calot's node (cysto-hepatic triangle or hepato-biliary triangle)	C772	Intra-abdominal	Para-aortic
Cardiac	C771	Intrathoracic	Mediastinal

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Cardial	C771	Intrathoracic	Mediastinal
Cardioesophageal	C771	Intrathoracic	Mediastinal
Carinal; tracheal bifurcation; tracheobronchial	C771	Intrathoracic	Mediastinal
Caval (para-)	C772	Intra-abdominal	Para-aortic
Cecal; anterior cecal: posterior cecal: prececal; retrocecal, NOS	C772	Intra-abdominal	Mesenteric
Celiac	C772	Intra-abdominal	Para-aortic
Central compartment (paralaryngeal, prelaryngeal [Delphian]) adjacent to thyroid gland.	C770	Head, face and neck	Cervical, right and left*
Cervical NOS, anterior deep cervical; deep cervical (scalene); lower deep cervical; upper/superior cervical; lower/inferior; middle deep cervical; posterior cervical (spinal accessory); transverse cervical (supraclavicular)	C770	Head, face and neck	Cervical, right and left*
Cloquet's node (inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Colic NOS, ileocolic, mesocolic, middle (right)	C772	Intra-abdominal	Mesenteric
Common bile duct	C772	Intra-abdominal	Para-aortic
Cubital	C773	Axilla or arm	Axillary, right and left*
Cystic duct	C772	Intra-abdominal	Para-aortic
Deep cervical (laterotracheal)	C771	Intrathoracic	Cervical, right and left*
Deep inguinal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Delphian node (prepharyngeal), adjacent to thyroid gland.	C770	Head, face and neck	Cervical, right and left*
Deltopectoral	C773	Axilla or arm	Axillary, right and left*
Diaphragmatic, sub	C771	Intrathoracic	Mediastinal
Duodenal	C772	Intra-abdominal	Para-aortic
Epicolic (Foramen of Winslow, omental)	C772	Intra-abdominal	Mesenteric
Epitrochlear	C773	Axilla or arm	Axillary, right and left*
Esophageal (para-, peri-)	C771	Intrathoracic	Mediastinal
Facial	C770	Head, face and neck	Cervical, right and left*
Femoral (superficial inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Foramen of Winslow (epicolic, omental)	C772	Intra-abdominal	Mesenteric
Gastric NOS, left (superior), gastrocolic; right (inferior gastric);	C772	Intra-abdominal	Mesenteric
Gastrocolic (Gastric)	C772	Intra-abdominal	Mesenteric
Gastroduodenal	C772	Intra-abdominal	Mesenteric
Gastroepiploic (gastro-omental)	C772	Intra-abdominal	Mesenteric
Gastrohepatic	C772	Intra-abdominal	Mesenteric
Gastro-omental (gastroepiploic)	C772	Intra-abdominal	Mesenteric
Gastropancreatic	C772	Intra-abdominal	Mesenteric
Gerota's node (promontorial, middle sacral)	C775	Pelvic	Para-aortic
Greater curvature	C772	Intra-abdominal	Mesenteric

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Greater omental	C772	Intra-abdominal	Mesenteric
Groin	C774	Inguinal region or leg	Inguino-femoral, right and left*
Hemorrhoidal NOS; inferior; middle; superior	C775	Pelvic	Pelvic, right and left*
Hepatic, NOS; hepatic artery; hepatic pedicle; hepatic, inferior vena cava; porta hepatis (hilar)	C772	Intra-abdominal	Para-aortic
Hepatoduodenal ligament	C772	Intra-abdominal	Para-aortic
Hilar (splenic)	C772	Intra-abdominal	Mesenteric
Hilar [in hilus of liver] (porta hepatis, portal)	C772	Intra-abdominal	Para-aortic
Hilar, bronchial	C771	Intrathoracic	Hilar, right and left*
Hilar, hepatoduodenal ligament	C772	Intra-abdominal	Para-aortic
Hilar, pulmonary root	C771	Intrathoracic	Hilar, right and left*
Hilar; bronchopulmonary, proximal lobar, pulmonary root	C771	Intrathoracic	Hilar, right and left*
Hypogastric (internal iliac)	C775	Pelvic	Pelvic, right and left*
Ileocolic	C772	Intra-abdominal	Mesenteric
Iliac, common	C775	Pelvic	Pelvic, right and left*
Iliac, external	C775	Pelvic	Pelvic, right and left*
Iliac, internal (hypogastric, obturator)	C775	Pelvic	Pelvic, right and left*
Inferior deep cervical (scalene)	C770	Head, face and neck	Cervical, right and left*
Inferior deep jugular	C770	Head, face and neck	Cervical, right and left*
Inferior gastric (right gastric)	C772	Intra-abdominal	Mesenteric
Inferior hemhorrhoidal	C775	Pelvic	Pelvic, right and left*
Inferior vena cava	C772	Intra-abdominal	Para-aortic
Infra-auricular	C770	Head, face and neck	Cervical, right and left*
Infraclavicular (subclavicular)	C773	Axilla or arm	Infraclavicular, right and left*
Infrapyloric (subpyloric)	C772	Intra-abdominal	Para-aortic
Inguinal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Inguinal NOS; deep, superficial (subinguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Innominate (thoracic)	C771	Intrathoracic	Mediastinal
Interaortocaval	C772	Intra-abdominal	Para-aortic
Intercostal	C771	Intrathoracic	Mediastinal
Interlobar (within the lung)/intrapulmonary	C771	Intrathoracic	Mediastinal
Internal iliac (hemorrhoidal)	C775	Pelvic	Pelvic, right and left*
Internal jugular	C770	Head, face and neck	Cervical, right and left*
Internal mammary (parasternal)	C771	Intrathoracic	Mediastinal
Interpectoral	C773	Axilla or arm	Axillary, right and left*
Intestinal	C772	Intra-abdominal	Mesenteric
Intra-abdominal	C772	Intra-abdominal	Mesenteric

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Intrabronchial, NOS	C771	Intrathoracic	Hilar
Intramammary lymph node	C773	Axilla or arm	Axillary, right and left*
Intrapancreatic lymph node	C772	Intra-abdominal	Para-aortic
Intraparotid	C770	Head, face and neck	Cervical, right and left*
Intrapelvic	C775	Pelvic	Pelvic, right and left*
Intrapulmonary (within the lung)	C771	Intrathoracic	Mediastinal
Intrapulmonary, segmental/subsegmental	C771	Intrathoracic	Mediastinal
Jugular, lower, mid, upper, internal	C770	Head, face and neck	Cervical, right and left*
Jugulodigastric (subdigastric)	C770	Head, face and neck	Cervical, right and left*
Jugulo-omohyoid (supraomohyoid)	C770	Head, face and neck	Cervical, right and left*
Lateral aortic (lumbar)	C772	Intra-abdominal	Para-aortic
Lateral compartment (jugular, mid and lower; supraclavicular; upper deep jugular; spinal accessory; retropharyngeal; submandibular; submental)l	C770	Head, face and neck	Cervical, right and left*
Lateral jugular	C770	Head, face and neck	Cervical, right and left*
Laterotracheal (anterior deep cervical)	C771	Intrathoracic	Cervical, right and left*
Left (superior) gastrocolic	C772	Intra-abdominal	Mesenteric
Leg/Lower limb	C774	Inguinal region or leg	Inguino-femoral, right and left*
Lesser curvature	C772	Intra-abdominal	Mesenteric
Lesser omental	C772	Intra-abdominal	Mesenteric
Lineal (splenic)	C772	Intra-abdominal	Mesenteric
Lobar, proximal (pulmonary)	C771	Intrathoracic	Hilar
Lobar/intrapulmonary	C771	Intrathoracic	Hilar
Lower jugular	C770	Head, face and neck	Cervical, right and left*
Lower peratracheal	C771	Intrathoracic	Mediastinal
Lower periesophageal (intrathoracic esophagus)	C771	Intrathoracic	Mediastinal
Lower peritracheal	C771	Intrathoracic	Mediastinal
Lumbar	C771	Intra-abdominal	Pelvis, right and left*
Lumbar aortic	C772	Intra-abdominal	Para-aortic
Mandibular	C770	Head, face and neck	Cervical, right and left*
Mastoid (postauricular, retro-auricular)	C770	Head, face and neck	Cervical, right and left*
Mediastinal, anterior	C771	Intrathoracic	Mediastinal
Mediastinal, NOS	C771	Intrathoracic	Mediastinal
Mediastinal, posterior (tracheoesophageal)	C771	Intrathoracic	Mediastinal
Mediastinal, superior	C771	Intrathoracic	Mediastinal
Mesenteric	C772	Intra-abdominal	Mesenteric
Mesenteric, inferior	C772	Intra-abdominal	Mesenteric

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Mesenteric, sigmoid	C772	Intra-abdominal	Mesenteric
Mesenteric, superior	C772	Intra-abdominal	Mesenteric
Mesocolic	C772	Intra-abdominal	Mesenteric
Mid jugular	C770	Head, face and neck	Cervical, right and left*
Midcolic	C772	Intra-abdominal	Pelvic, right and left*
Middle (right) colic	C772	Intra-abdominal	Mesenteric
Middle hemhorrhoidal	C775	Pelvic	Pelvic, right and left*
Middle sacral	C775	Pelvic	Pelvic, right and left*
Nasolabial	C770	Head, face and neck	Cervical, right and left*
Node of Cloquet's or Rosenmuller (highest deep inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Obturator	C775	Pelvic	Pelvic, right and left*
Occipital; suboccipital	C770	Head, face and neck	Cervical, right and left*
Omental	C772	Intra-abdominal	Mesenteric
Pancreatic; Aselli's glands (nodes near pancreas); parapancreatic; peripancreatic	C772	Intra-abdominal	Para-aortic
Pancreaticoduodenal	C772	Intra-abdominal	Para-aortic
Pancreaticosplenic (pancreaticolineal)	C772	Intra-abdominal	Mesenteric
Para-aortic	C772	Intra-abdominal	Para-aortic
Parabronchial	C771	Intrathoracic	Mediastinal
Paracardial	C772	Intra-abdominal	Mesenteric
Paracaval	C772	Intra-abdominal	Para-aortic
Paracervical	C775	Pelvic	Pelvic, right and left*
Paracolic/pericolic	C772	Intra-abdominal	Para-aortic
Paraesophageal	C771	Intrathoracic	Mediastinal
Paralaryngeal	C770	Head, face and neck	Cervical, right and left*
Parametrial	C775	Pelvic	Pelvic, right and left*
Parapancreatic	C772	Intra-abdominal	Para-aortic
Parapharyngeal	C770	Head, face and neck	Cervical, right and left*
Parasternal (internal mammary)	C771	Intrathoracic	Mediastinal
Paratracheal	C771	Intrathoracic	Mediastinal
Paratracheal, lower	C771	Intrathoracic	Mediastinal
Parotid (peri-)	C770	Head, face and neck	Cervical, right and left*
Pectoral (anterior axillary)	C773	Axilla or arm	Axillary, right and left*
Pelvic, NOS	C775	Pelvic	Pelvic, right and left*
Peratracheal, lower	C771	Intrathoracic	Mediastinal
Peri-aortic	C772	Intra-abdominal	Para-aortic

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Peribronchial; parabronchial	C771	Intrathoracic	Mediastinal
Pericardial	C771	Intrathoracic	Mediastinal
Pericaval	C772	Intra-abdominal	Para-aortic
Pericholedochal	C772	Intra-abdominal	Para-aortic
Pericolic	C772	Intra-abdominal	Mesenteric
Periduodenal	C772	Intra-abdominal	Para-aortic
Periesophageal	C771	Intrathoracic	Mediastinal
Perigastric, except cardiac	C772	Intra-abdominal	Mesenteric
Peripancreatic	C772	Intra-abdominal	Para-aortic
Periparotid	C770	Head, face and neck	Cervical, right and left*
Periportal	C772	Intra-abdominal	Pelvic, right and left*
Periprostatic	C775	Pelvic	Pelvic, right and left*
Perirectal	C775	Pelvic	Pelvic, right and left*
Peritracheal, lower	C771	Intrathoracic	Mediastinal
Periureteral	C772	Intra-abdominal	Para-aortic
Perivesical	C775	Pelvic	Pelvic, right and left*
Pharyngeal; Delphian node; prepharyngeal; retropharyngeal	C770	Head, face and neck	Cervical, right and left*
Phrenic; inferior phrenic vein; superior phrenic vein	C771	Intra-thoracic	Mediastinal
Popliteal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Porta hepatis [in hilus of liver]	C772	Intra-abdominal	Para-aortic
Portal (portal vein)	C772	Intra-abdominal	Para-aortic
Postauricular	C770	Head, face and neck	Cervical, right and left*
Posterior axillary	C773	Axilla or arm	Axillary, right and left*
Posterior cecal	C772	Intra-abdominal	Para-aortic
Posterior mediastinal	C771	Intrathoracic	Mediastinal
Posterior triangle (spinal accessory and transverse cervical)	C770	Head, face and neck	Cervical, right and left*
Preaortic	C772	Intra-abdominal	Para-aortic
Pre-auricular	C770	Head, face and neck	Cervical, right and left*
Precarinal	C771	Intrathoracic	Mediastinal
Prececal	C772	Intra-abdominal	Mesenteric
Prelaryngeal	C770	Head, face and neck	Cervical, right and left*
Prepharyngeal	C770	Head, face and neck	Cervical, right and left*
Presymphsial	C775	Pelvic	Pelvic, right and left*
Pretracheal	C770	Head, face and neck	Cervical, right and left*
Promontorial	C775	Pelvic	Pelvic, right and left*
Proximal lobar	C771	Intrathoracic	Hilar

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Proximal mesentery	C772	Intra-abdominal	Mesenteric
Pulmonary ligament	C771	Intrathoracic	Mediastinal
Pulmonary root	C771	Intrathoracic	Hilar
Pulmonary, NOS	C771	Intrathoracic	Hilar
Pyloric, Infra (subpyloric)	C772	Intra-abdominal	Para-aortic
Pyloric; suprapyloric	C772	Intra-abdominal	Para-aortic
Rectal	C775	Pelvic	Pelvic, right and left*
Recurrent laryngeal (laterotracheal, anterior deep cervical)	C770	Head, face and neck	Cervical, right and left*
Recurrent pharyngeal	C770	Head, face and neck	Cervical, right and left*
Renal hilar	C772	Intra-abdominal	Para-aortic
Retroaortic	C772	Intra-abdominal	Para-aortic
Retroauricular (mastoid)	C770	Head, face and neck	Cervical, right and left*
Retrocaval	C772	Intra-abdominal	Para-aortic
Retrocecal	C772	Intra-abdominal	Para-aortic
Retrocecal (posterior cecal)	C772	Intra-abdominal	Para-aortic
Retrocrural	C771	Intra-thoracic	Mediastinal
Retroperitoneal	C772	Intra-abdominal	Para-aortic
Retropharyngeal	C770	Head, face and neck	Cervical, right and left*
Retrotracheal	C771	Intrathoracic	Mediastinal
Right (inferior) gastric	C772	Intra-abdominal	Mesenteric
Rosenmuller's node (inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Rosenmuller's-Cloquet's nodes (inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Rotter's nodes (interpectoral between major and minor pectoralis)	C773	Axilla or arm	Axillary, right and left*
Rouviere's node (retropharyngeal)	C770	Head, face and neck	Cervical, right and left*
Sacral, lateral (laterosacral)	C775	Pelvic	Pelvic, right and left*
Sacral, middle (promontorial/promontory) (Gerota's node)	C775	Pelvic	Pelvic, right and left*
Sacral, NOS	C775	Pelvic	Pelvic, right and left*
Sacral, presacral	C775	Pelvic	Pelvic, right and left*
Sacral, uterosacral	C774	Pelvic	Pelvic, right and left*
Scalene (inferior deep cervical)	C770	Head, face and neck	Cervical, right and left*
Sigmoidal (sigmoid mesenteric)	C772	Intra-abdominal	Mesenteric
Spermatic vein	C774	Inguinal region or leg	Inguino-femoral, right and left*
Spinal accessory (posterior cervical)	C770	Head, face and neck	Cervical, right and left*
Splenic (hilar)	C772	Intra-abdominal	Mesenteric
Splenic (lienal)	C772	Intra-abdominal	Mesenteric
Subaortic (aortico-pulmonary window)	C772	Intra-abdominal	Mediastinal
Subcarinal	C771	Intrathoracic	Mediastinal

Lymph Node/Lymph Node Chain	Use for MP/H	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Subclavian (apical)	C770	Head, face and neck	Cervical, right and left*
Subclavicular (infraclavicular)	C773	Axilla or arm	Infraclavicular, right and left*
Subdigastric	C770	Head, face and neck	Cervical, right and left*
Subinguinal (superficial inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Sublingual	C770	Head, face and neck	Cervical, right and left*
Submandibular (submaxillary)	C770	Head, face and neck	Cervical, right and left*
Submental	C770	Head, face and neck	Cervical, right and left*
Suboccipital	C770	Head, face and neck	Cervical, right and left*
Subpleural (in the periphery of the lung)	C771	Intrathoracic	Mediastinal
Subpyloric	C772	Intra-abdominal	Para-aortic
Subscapular (posterior axillary)	C773	Axilla or arm	Axillary, right and left*
Superficial inguinal (subinguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Superior gastrocolic (left gastrocolic)	C772	Intra-abdominal	Mesenteric
Superior hemhorrhoidal	C775	Pelvic	Pelvic, right and left*
Superior hilum	C772	Intra-abdominal	Pelvic, right and left*
Superior jugular	C770	Head, face and neck	Cervical, right and left*
Superior mesenteric	C772	Intra-abdominal	Pelvic, right and left*
Superior rectal	C775	Pelvic	Pelvic, right and left*
Supraclavicular (transverse cervical)	C770	Head, face and neck	Cervical, right and left*
Supraomohyoid	C770	Head, face and neck	Cervical, right and left*
Suprapancreatic	C772	Intra-abdominal	Para-aortic
Suprapyloric	C772	Intra-abdominal	Para-aortic
Thoracic (innominate)	C771	Intrathoracic	Mediastinal
Thyroid	C770	Head, face and neck	Cervical, right and left*
Tibial	C774	Inguinal region or leg	Inguino-femoral, right and left*
Tracheal bifurcation	C771	Intrathoracic	Mediastinal
Tracheal; pretracheal; retrotracheal	C771	Intrathoracic	Mediastinal
Tracheobronchial	C771	Intrathoracic	Mediastinal
Tracheoesophageal (posterior mediastinal)	C771	Intrathoracic	Mediastinal
Transverse cervical (supraclavicular)	C770	Head, face and neck	Cervical, right and left*
Transverse cervical; posterior triangle; spinal accessory	C770	Head, face and neck	Cervical, right and left*
Trosier's node (left supraclavicular)	C770	Head, face, and neck	Cervical, right and left*
Upper jugular	C770	Head, face and neck	Cervical, right and left*
Virchow's node (supraclavicular)	C770	Head, face, and neck	Cervical, right and left*

*The right and left are separate regions per AJCC

Appendix D
New Histology Terms and Codes Hematopoietic and Lymphoid Neoplasms

Table D1a: New Histology Terms and Codes – Alphabetic List

Table D1a contains an alphabetic list of hematopoietic and lymphoid neoplasm histology codes and terms documented in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Ed. published in 2008. Use this table to code the histology when any of these more specific terms are the diagnosis. Column 1 is the more specific histology term; column 2 is the new code WHO proposed for that specific histology. These neoplasms are not newly reportable; they are more specific terms for diseases that would otherwise be coded in NOS categories. Do not use these codes for neoplasms diagnosed prior to 2010. The new codes will go into effect with cases diagnosed 1/1/2010 and after. There are no plans or mandates to identify 2008 and 2009 cases to recode using these codes.

New Histology Term	ICD-O Code
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); RPN1EVI1	9869/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3
ALK positive large B-cell lymphoma	9737/3
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A PBX1 (TCF3 PBX1)	9818/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3
Fibroblastic reticular cell tumor	9759/3
Hydroa vacciniforme-like lymphoma	9725/3
Intravascular large B-cell lymphoma	9712/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged	9807/3
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Myeloid neoplasms with PDGFRB rearrangement	9966/3
Myeloid and lymphoid neoplasms with PDGFRA rearrangement	9965/3
Myeloid leukemia associated with Down Syndrome	9898/3
Plasmablastic lymphoma	9735/3

New Histology Term	ICD-O Code
Polymorphic PTLD	9971/3
Primary cutaneous follicle center lymphoma	9597/3
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3

Table D1b: New Histology Terms and Codes – Numeric List

Table D1b contains a numeric list of hematopoietic and lymphoid neoplasm histology codes and terms documented in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Ed. published in 2008. Use this table to code the histology when any of these more specific terms are the diagnosis. Column 1 is the more specific histology term; column 2 is the new code WHO proposed for that specific histology. These neoplasms are not newly reportable; they are more specific terms for diseases that would otherwise be coded in NOS categories. Do not use these codes for neoplasms diagnosed prior to 2010. The new codes will go into effect with cases diagnosed 1/1/2010 and after. There are no plans or mandates to identify 2008 and 2009 cases to recode using these codes.

New Histology Term	ICD-O Code
Primary cutaneous follicle center lymphoma	9597/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3
Intravascular large B-cell lymphoma	9712/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
Hydroa vacciniforme-like lymphoma	9725/3
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Plasmablastic lymphoma	9735/3
ALK positive large B-cell lymphoma	9737/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Fibroblastic reticular cell tumor	9759/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged	9807/3
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3

New Histology Term	ICD-O Code
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A PBX1 (TCF3 PBX1)	9818/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3
Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); RPN1EVI1	9869/3
Myeloid leukemia associated with Down Syndrome	9898/3
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Myeloid and lymphoid neoplasms with PDGFRA rearrangement	9965/3
Myeloid neoplasms with PDGFRB rearrangement	9966/3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Polymorphic PTLD	9971/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3

Table D2: Histologic Terms and Codes with Changes in Case Reportability (Newly Reportable Conditions)

Table D2 contains hematopoietic and lymphoid neoplasms with changes in behavior from /1 (borderline malignancy) to /3 (malignant). The changes in histology codes and terms are documented in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition published in 2008. Reporting these neoplasms will go into effect with cases diagnosed 1/1/2010 and after. There are no plans or mandates to collect 2008 and 2009 cases having these diagnoses.

Histology Term	ICD-O Code
Langerhans cell histiocytosis, NOS	9751/3
T-cell large granular lymphocytic leukemia / Chronic lymphoproliferative disorder of NK-cells	9831/3
Myeloproliferative neoplasm, unclassifiable / Myelodysplastic/Myeloproliferative neoplasm, unclassifiable	9975/3

Appendix E
2010 and 2012 Obsolete Hematopoietic Neoplasm Codes

The table below is based on the 2008 WHO (see Page 12 of this manual). The table lists the obsolete ICD-O-3 codes, the obsolete descriptions, the effective obsolete dates, the current ICD-O-3 codes (per the Hematopoietic database), and the current descriptions.

OBS ICD-O-3 Code	Obsolete Description	Effective Obsolete Date	Current ICD-O-3 Code	Current Description
9654/3	Hodgkin lymphoma, lymphocytic depletion, diffuse fibrosis	1/1/2010	9653/3	Lymphocyte-depleted classical Hodgkin lymphoma
9661/3	Hodgkin granuloma	1/1/2010	9650/3	Hodgkin lymphoma, NOS
9662/3	Hodgkin sarcoma	1/1/2010	9650/3	Hodgkin lymphoma, NOS
9664/3	Hodgkin lymphoma, nodular sclerosis, cellular phase	1/1/2010	9663/3	Nodular sclerosis classical Hodgkin lymphoma
9665/3	Hodgkin lymphoma, nodular sclerosis, grade 1	1/1/2010	9663/3	Nodular sclerosis classical Hodgkin lymphoma
9667/3	Hodgkin lymphoma, nodular sclerosis, grade 2	1/1/2010	9663/3	Nodular sclerosis classical Hodgkin lymphoma
9670/3	Malignant lymphoma, small B lymphocytic, NOS	1/1/2012	9823/3	CLL/small lymphocytic lymphoma
9675/3	Malignant lymphoma, mixed small and large cell, diffuse	1/1/2010	9690/3	Follicular lymphoma
9684/3	Malignant lymphoma, large B-cell, diffuse, immunoblastic, NOS	1/1/2012	9680/3	Diffuse large B-cell lymphoma (DLBCL), NOS
9728/3	Precursor B-cell lymphoblastic lymphoma	1/1/2010	9811/3	B lymphoblastic leukemia/lymphoma, NOS
9729/3	Precursor T-cell lymphoblastic lymphoma	1/1/2010	9837/3	T lymphoblastic leukemia/lymphoma
9733/3	Plasma cell leukemia	1/1/2010	9732/3	Plasma cell myeloma
9750/3	Malignant histiocytosis	1/1/2010	9751/3	Langerhans cell histiocytosis
9752/1	Langerhans cell histiocytosis, unifocal	1/1/2010	9751/3	Langerhans cell histiocytosis
9753/1	Langerhans cell histiocytosis, multifocal	1/1/2010	9751/3	Langerhans cell histiocytosis
9754/3	Langerhans cell histiocytosis, disseminated	1/1/2010	9751/3	Langerhans cell histiocytosis
9760/3	Immunoproliferative disease, NOS	1/1/2010	9761/3 9762/3	Code to more specific immunoproliferative disease. See codes 9761/3 and 9762/3
9764/3	Immunoproliferative small intestinal disease	1/1/2010	9762/3	Alpha heavy chain disease
9805/3	Acute biphenotypic leukemia	1/1/2010	9806/3-9809/3	Assign to one of the new codes in the 9806-9809 range. For acute biphenotypic leukemia, NOS, assign code: 9809/3
9835/3	Precursor cell lymphoblastic leukemia, NOS	1/1/2010	9811/3	B lymphoblastic leukemia/lymphoma, NOS
9836/3	Precursor B-cell lymphoblastic leukemia/lymphoma	1/1/2010	9811/3	B lymphoblastic leukemia/lymphoma, NOS
9960/3	Chronic myeloproliferative disease	1/1/2010	9975/3	Myelodysplastic/myeloproliferative neoplasm unclassifiable
9984/3	Refractory anemia with excel blasts in transformation	1/1/2010	9983/3	Refractory anemia with excess blasts
9987/3	Therapy related myelodysplastic syndrome	1/1/2010	9920/3	Therapy-related myeloid neoplasm